

PEDIATRICS

Handwritten Note

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Name: _____

Subject: _____ **PEDIATRICS**



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PEDIATRICS

Neonate - First 4 wk. of life.

- ↳ Early - 7 days, cause of death - Prematurity
- Late - 7-28 days, - Sepsis.

Term - 37-42 wk gestation

Preterm - < 37 wk

Post term - > 42 wk

Normal wt \rightarrow 2.5-4 kg

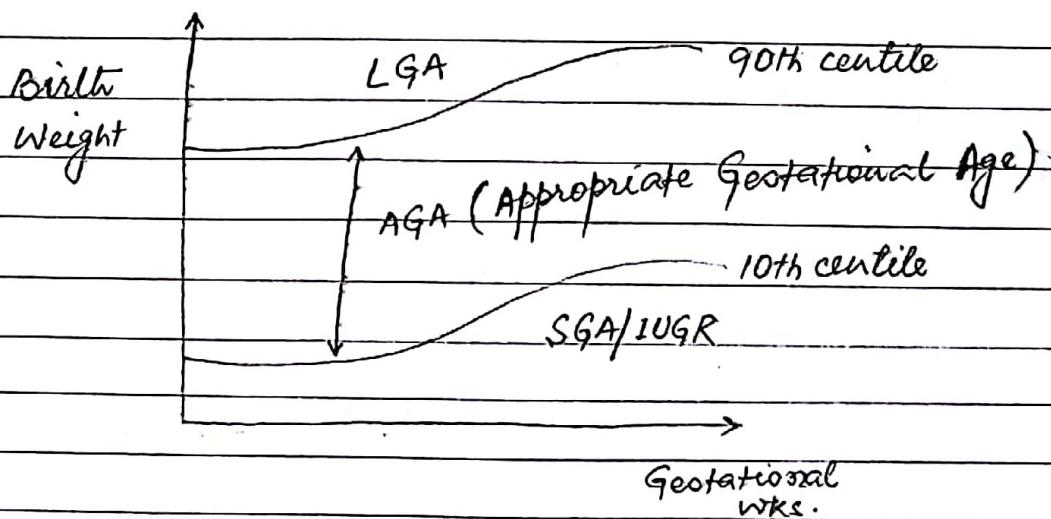
LBW < 2.5 kg

VLBW < 1.5 kg

ELBW < 1 kg

Macrosomia $>$ 4 kg.

Lubchencho chart:



Constitutional:

- LGA (Large Gestational Age)
- Short stature.
- Delayed puberty.

- All term baby has 6 fontanelle:
 - Anterior → ~~closed~~
 - Posterior
 - 2 sphenoid
 - 2 Mastoid.

Posterior fontanel - closes at birth.
open in 3% baby.

Anterior fontanel - diamond shaped.

2.5 X 2.5 cms.

At level

Pulsatile

closes at 18-24 months of life.

Craniosynostosis:

^{all}
Early closure of fontanel.

Complication:

- Microcephaly
- IGT ↑ → optic atrophy.
- cosmetic.

Rx - Cranieotomy

Syndromes associated - Apert
Crouzon] AD
Pfeiffer
Carpenter - AR

Delayed closure of fontanel:

- Rickets
- Down's
- Hypothyroidism
- Hypophosphatasia
- cleidocranial dysostosis.

Q. 2 wks baby, Hypotonia, hypothermia, umbilical hernia, constipation, physiological jaundice prolonged.
 ↳ Congenital hypothyroidism
 (humeral head - Epiphyseal dysgenesis)
 (46④ epiphysis of bone)

Neonatal Screening:

- TSH; T_4
 - Delayed rise of TSH
 - TBG deficiency (TSH↑; $T_4 \downarrow$)
 - Best time: after 48 hrs (72 hrs)

M/c/c of congenital hypothyroidism - 85% agensis/dysgenesis of thyroid gland.

Congenital hypothyroidism:

Prevalance = 1: 2000

Girls > Boys.

Neonatal screening : Continue

TSH; T_4

- Cold temp^r
- Physiological TSH surge 48 hrs.
- APP > 48 hrs to 6 days.
- In OP Ghai → 3-5 days



→ Sample obtained → Heel prick.
 Safe area → Side prick.

- pt in shock, venous access can't get in 60 sec.
- M/c^{other} site - Near tibial tuberosity.
 Upper end of fibia
 Lower end of femur.
- In shock → i.v. fluids → 20 ml/kg bodies NS.

Most easily accessible venous route - Umbilical Vein.

Phenylketonuria: Deficiency of Phenylalanine hydroxylase.

Phenylalanine

PKU \ominus ~~PKU~~ $\not\rightarrow$ Phenylalanine hydroxylase.

~~Phenylalanine~~

Tyrosine

\downarrow Tyrosinase

DOPA

\downarrow

Dopa quinone

\downarrow

Melanin

\uparrow Phenylalanine \rightarrow Toxic to Brain \rightarrow so in PKU High Phenylalanine

- Child Mental Retardation.

20 mg/dl

- Developmental delay.

- Exaggerated Reflexes

- Microcephaly.

Rx - PKU

- Supplement Tyrosine
- Restrict Phenylalanine
- Now Tyrosine essential.
- Aim: Serum Phenylalanine $< 6 \text{ mg/dl}$.
- Lifelong.

Q. About PKU Rx, first step -

A) Stop the substrate of the enzyme

B) Supplement the enzyme.

C) Reduce the substrate of the enzyme

D) Provide deficient protein.



Date

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Maple Syrup Urine disease (MSUD):

- Deficiency of α -keto acid branched chain dehydrogenase.
- High Valine : Leucine : Isoleucine Blood & CSF.
- Coma
- Severe acidosis.
- We do dialysis - To remove Valine : Lev : Isoleucine.

Neonatal Screening : Conclusion

- Tandem mass
 - Spectrophotometry \rightarrow metabolic
- Cystic fibrosis
- Congenital adrenal hyperplasia
- G6PD deficiency.
- Biotin deficiency.

ANTHROPOMETRY : TERM BABY

Length - 50 cms

Head circumference - 35 cm

$HC > CC$ but not more than 3 cm.

- $HC > CC (> 3\text{cm}) \rightarrow$ Congenital hydrocephalus.
 \rightarrow Asymmetric IUGR baby.

$CC = HC \rightarrow$ at 9-12 months

By 1 yr ; $CC \ggg HC$

Upper segment : Lower segment (US : LS)

New born = 1.7 : 1

At 10 yrs = 1 : 1

Adult = 0.9 : 1

Achondroplasia : Short limb dwarf (US/LS \uparrow)

Hypothyroidism : US/LS \uparrow (disproportionate short stature)

Meconium:

95% pass meconium at 24 hrs.

99% " " " 48 hrs.

Cause of delayed passage meconium

- Imperforate anus

- Hirschsprung / aganglionic
 ↳ Rectum biopsy.

- Meconium ileus → Cystic fibrosis.

- ↳ Small intestinal obstruction.

Q 48 hrs baby has not passed meconium. Next Ix

A) CFTR gene test

B) Sweat chloride

C) Manometry

✓ D) Lower GI contrast study.

 ↳ △ Hirschsprung

Treat meconium ileus.

Delayed Urine:

- B/L Renal agenesis

- ↳ Maternal oligohydramnios.

Post. Urethral valve

also cause

B/L pulm. hypoplasia



POTTER'S Sequence.

Potter's face - Nose pinched in

Retrognathia

Micrognathia

Bag & mask → Pneumothorax.

Q If a baby has not passed urine in 1st 48 hrs, Next Ix
→ USG

Q 3 days old, c/o - Weak dribbling urine stream.

O/E - palpable distended bladder.

Δ → Posterior Urethral valve.

IOC : MCU (Micturating Cysto Urethrography).

Rx : Cystoscopic Fulguration

SGA / IUGR :

Complication : TORCH infection / Chromosomal disorders.

Symptom - Asphyxia → causes MAS

↓ (Meconium Aspirated Syndrome)

PPHN (Persistent pulmonary HTN of Newborn).

Severe IUGR - Pulmonary hemorrhage

Limited stores - Hypoglycemia, HypoCa; HypoMg.

- Polycythemia

↳ bcoz in IUGR erythropoietin is very sensitive to hypoxia.

- Neutropenia

- Thrombocytopenia.

Q Full term small for date babies are more disposed to -

a) HyperCa

b) CNS infection

c) PDA

common in preterm.

✓ d) Hypoglycemia

Symmetric IUGR

Asymmetric IUGR

Cause	Chromosomal/ Torch	Maternal Complications
Cell no.	↓	(N)
Cell size	↓	↓
Brain	↓	(N) spared. HC > CC > 3cm Brain/liver ↑

PONDERAL'S INDEX:

$$\frac{wt \text{ (gm)}}{Length \text{ (cm)}^3} \times 100$$

< 2 → Asymmetric IUGR

≥ 2 → AGA / Symmetric IUGR

Normal Neonatal phenomena:

Milia: Distended Sebaceous gland on face & nose.

Erythema Tonicum: Erythema on face & trunk
2-3 days of life.

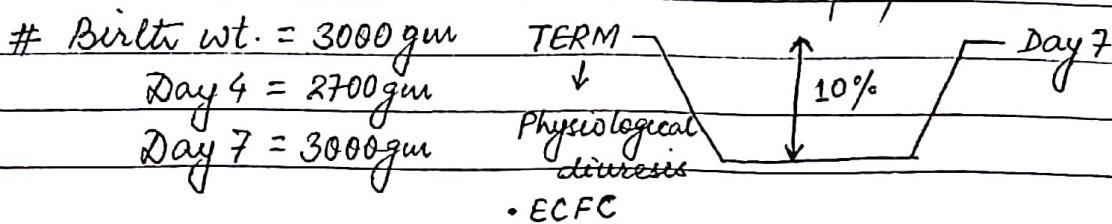
Stork bites: Pinkish gray capillary hemangioma
on back & buttocks.

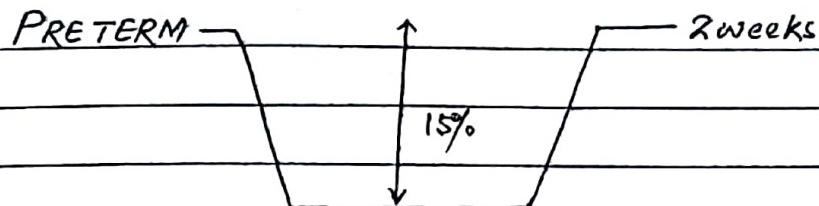
Epstein Pearl: Epithelial inclusion cyst on palate
& prepuce.

Natal teeth (Pre deciduous teeth): lower incisor position.

~~Withdrawl~~ Withdrawl vaginal bleeding: On 5th - 7th day.

Acrocyanosis (Peripheral Cyanosis): Limbs cyanosed
Lips pink.





IUGR → do not lose wt (ECF compact)
 ↳ wt. stable for 1-2 days then wt. gain.

Q. Not normal in a newborn?

- A) Proteinuria] in ELBW
- B) Glucosuria]
- C) 1-2 pus cells/hpf
- D) Bacteriuria

Neonatal Reflexes:

① Moro Reflex:

1st phase - Abduction of shoulder joint
 Extension of elbow joint.
 opening of fingers

2nd phase - Adduction & flexion.

- Appears 28-32 wks gestation.

• Adduction / complete → 36-38 wks.

- Disappears at around 2-3 months of life.

- Persistence beyond 6 months abnormal.



Cerebral palsy.

≡ - Asymmetric moro's

- Brachial plexus injury.
- # clavicle
- # Humerus
- Hemiplegia.

Early hand preference is always abnormal

- Hemiplegia at other side.

- 95% cases → Hemiplegia at Rt. side.

Exaggerated Moro → HIE - 1

ATNR (Asymmetric Tonic Neck Reflex):

Side of face - Extended

Side of occiput - Flexed.

Onset - 35 wks

Fully developed - 1 month

Duration - 6-7 months

Do not ~~roll~~ roll (Rolling start when this reflex disappears)

Disappears - 6 months

STNR (Symmetric tonic neck reflex):

Neck extended

→ Zone ↑ UL

↓ LL

Neck flexed

↓

Zone ↓ in all limbs.

- Not present at birth.

- Appears 4-6 months of life.

- Disappears 8-12 months of life.

- Child starts to crawl when this reflex disappears.

Parachute reflex:

- Not at birth

- Appears 6-7 months of life.

- Well developed at 10-11 months of life.
- Persists life long.

LANDAU Reflex:

- Appears at 3 months of life
- Disappears at 1 year of life.

On ventral suspension - spine strengthens / straightens.

Child gets out of flexion attitude by this reflex.

GRASP Reflex:

- Appears 28 wks of gestation
- Well developed 32 wks
- Disappears 3 months of life.

Sucking & Rooting reflex:

< 28 wks absent.

At 28 wks - Some sucking bursts.

32 wks - Appear

34 wks - Co-ordination

Q

31 wks ; 1500gms ; Feed - ?

A) Enteral → NG tube (Expressed Breast Milk)

B) Enteral + i.v. fluids

C) IV fluids.

D) TPN.

Q

33 wks ; 1500gms ; Feed - ?

↳ Enteral → Katori / Paladay / Spoon

(Expressed Breast Milk)



AIIMS NICU Protocol 2014 :

> 34 wks - Breast feed

32-34 wks - Katori (Expressed breast milk)

< 32 wks - NG tube (" " " ")

< 1200gms - i.v. dextrose + minimal enteral
feeds 10-15 ml/kg/day.Rapid enteral feed can cause - Necrotising
Enterocolitis.

Fetal alcohol Syndrome :

- Skin folds at the corner of the eyes
- Low nasal bridge.
- Short nose
- Indistinct philtrum (groove b/w nose & upper lip)
- Small head circumference (microcephaly)
- Small eye opening!
- Flat mid face / midfacial or maxillary hypoplasia.
- Thin upper lip.
- Septal defects < ASD
VSD.

Q Ass. C ~~Ans~~ fetal alcohol Syndrome except -

- A) Microcephaly
- B) Overgrowth
- C) Flat face
- D) Small palpebral fissure.

PREMATURITY

Respiratory System : RDS (Respiratory distress Syndrome).

Chronic lung disease /

Bronchopulmonary dysplasia

O₂ dependence on
4 wk of life

Rx : Home O₂ therapy.

Prevention : Nasal CPAP.

Vit A supplementation.

Furosemide

Keep underhydrated in ICU.

CNS : ① Apnea (> 20 sec); or any period if associated central cyanosis & bradycardia.

Q M/c signs of acute hypoxia in neonates

A) Bradycardia

B) Tachycardia.

Types of Apnea : 3 types

① Central : asphyxia ; preterm.

② Obstructive :

all newborn are obligate nose breathers till
4 months of life.

Q Full term newborn

episodes of cyanosis - worsen when feed.

Seems better when crying.

↳ Choanal atresia → B/L , posterior.

③ Mixed (M/c)



Apnea of prematurity:

Risk: < 28 weeks $\rightarrow 100\%$

Onset: 1-2 days; never > 7 days.

Rx :

1st step: Nasal CPAP

Methylxanthines - Aminophylline (^{Narrow therapeutic range})
Caffeine Cilrāte (DOC)
 \hookrightarrow wide margin of safety

L Loading dose of Aminophylline: 5-6 mg/kg.
followed by maintenance dose
1-2 mg/kg every 6-8 hrs.

Caffeine Cilrāte: Loading dose \rightarrow 20 mg/kg

Maintenance dose \rightarrow 1-5 mg/kg/day

(2) Intracranial hemorrhage

(Only subdural hemorrhage is common in term baby).

- Capillaries in the subependymal germinal matrix is fragile; so they rupture.

Preterm; sudden pale; shock, fontanel bulging; seizures - Intra ventricular hemorrhage (IVH)



Risk IVH preterms - < 1500 gm $\rightarrow 30\%$

[50% IVH occurs in 24 hrs.

[75% " " 72 hrs.

IOC for newborn having seizure \rightarrow Trans fontanel USG

Term; Breech \rightarrow IVH

Instrumental delivery \rightarrow IVH

Prevention of IVH:

- Antenatal steroids
- Low dose indomethacin to baby.
- Prevent acidosis, infections in baby.

③ ~~Asphyxia~~ Asphyxia

Preterm

Term

Periventricular
leucomalacia (PVL)

Parasagittal injury

Power: UL > LL

Spastic quadriplegic cerebral
palsy.

Diplegia

(upper limbs strong
LL are weak)

↳ Spastic diplegia type
of Cerebral palsy.

MRI: Coronal Section

- parasagittal injury.

• Mentally Retarded

Q.

M/c sequel of Periventricular
leucomalacia in preterm

- Spastic diplegia

MRI: PVL (IOC)

- less white matter

- Shrinkage Ventricle.

- Mentally Retarded.



Status marmoratus: diffuse neuronal loss following asphyxia.

APGAR Score : 1, 5, 10 minutes

- doesn't help in resuscitation.
- 5'; 7'; 10' low → Worse neuronal outcome.

0 1 : 2

A = Appearance Central cyanosis / Acrocyanosis Pink.
Pale

P = Pulse (HR) 0 <100 >100

G = Grimace No Grimace Crying

A = Attitude Extended Mid Flexed

R = Respir^r effort Apnea Gasping Crying.

NORMAL - 7-10

Moderate to severe asphyxia ; out of hospital CPR

↓
ischemia reperfusion injury

↓
Free radical damage.

Rx - Therapeutic hypothermia / Selective head cooling:
33.5°C in 6 hrs of life ; keep for 72 hrs!

↓
preventive

Hypoxic Ischemic Encephalopathy (HIE):

Injury to brain at severe asphyxia.

Sigors	STAGE 1	STAGE 2	STAGE 3
• Loss of consciousness	<u>Hyperalert</u>	<u>Lethargic</u>	<u>Stuporous; Coma</u>
• Muscle tone	Normal	Hypotonic	<u>Flaccid</u>
• Posture	Normal	Flexion	<u>Decerebrate</u>
• Tendon reflexes/ clonus	<u>Hyperactive</u>	<u>Hyperactive</u>	Absent
• Myoclonus	+nt	+nt	-nt
• Moro reflex	<u>Strong</u>	<u>Weak</u>	-nt
• Pupils	<u>Mydriasis</u>	<u>Miosis</u>	<u>Unequal, poor light reflex.</u>
• Seizures	<u>None</u>	<u>Common</u>	<u>Decerebration</u>
• EEG finding	<u>Normal</u>	<u>Low voltage changing to seizure activity.</u>	<u>Burst suppression to isoelectric</u>
• Duration	< 24 hr if progresses; otherwise may remain	24 hrs - 14 days	Days to weeks
• Outcome	Good.	Variable	Death, Severe deficit

DOC: Seizures in Newborn: Phenobarbitone.

(Bolus, 20 mg/kg)



③ CVS: Hypotension; PDA (Patent ductus arteriosus).

PDA: Preterm \Rightarrow Asphyxia \rightarrow PGs

Term \Rightarrow Rubella infection \rightarrow Vessel wall defect.

\downarrow
Rx: Surgery

In preterm: Rx: NSAIDs

(Ibuprofen $>$ Indomethacin)
 \downarrow

Less nephrotoxic

If Medical management fails \rightarrow Sx

C/F of PDA:

- 6-10 wks of life CHF
- Preterm baby, failure to wean off ventilator (hypoxia; CO₂ retention)

O/E: • Bounding pulses \bar{c} wide pulse pressure.

• Continuous machinery murmur at the upper left sternal border.

④ GI System: Necrotising Enterocolitis

⑤ Eye: ROP/ Retrolental fibroplasia.

⑥ Hypothermia; ^{Hypo}Glycaemia; HypoCa; HypoMg.

⑦ Anemia; jaundice; Infection.



Necrotizing Enterocolitis (NEC) :

R/F: ① Immature Gut.

NEC: Susceptibility of premature infants

- Reduced proteolytic enzymes
 - ↑ Gastric pH
 - ↓ peristalsis
 - ↓ motility
 - Altered epithelial membr & tight junction
 - Altered bacterial flora.
 - ↓ mucous coat
 - Altered mucous protein
 - ↑ epithelial permeability.
- Sepsis toxins
 - Top fed (cow milk)

② Mature cocaine

③ PPIs ; Anti H₂

④ Rapid advancement of feed.

Prevention : ① Antenatal steroids

- ↓ IVH
- ↓ PVL
- ↓ NEC
- ↓ RDS
- ↓ Neonatal mortality.

HUMAN MILK : ↑ proteolytic enzyme

↓ Gastric pH

↑ peristalsis

↑ motility

less pathogenic bacterial flora



- Altered mucus coat (Improves)
- ↓ epithelial permeability.

(2) Trophic feeds - expressed milk
10-15 ml/kg/day.

(3) Avoid PPIs; anti H₂

(4) Avoid rapidly feed advance.

(5) Probiotics.

Q. 1 yr old infant; 10-12 episodes of watery stools / day for last 9 days. Along c Zn, which else should be advised -

A) ORS c antibiotics

B) ORS orally.

C) ORS c low lactose diet

D) ORS c low lactose diets & probiotics.

PROBIOTICS:

Prevents NEC - *Lactobacillus acidophilus*
& *Bifidobacterium infantis*
to VLBW's

- may use in Rotavirus:

Lactobacillus rhamnosus
& *Saccharomyces boulardii*

Modified Bell's Staging for NEC:

Stage Ia:	Suspected NEC	Distension; ileus; occult blood in stool
I _b :	Suspected	Gross blood loss.
IIa:	Definite	Focal pneumoperitoneum
II _b :	Definite	HMT; Diffuse pneumoperitoneum; Portal Venous Gas.
IIIa:	Advanced	DIC; Shock; Peritonitis
III _b :	Advanced-perforation	Pneumoperitoneum

Q. Neonate; distended abdomen & B/L gas shadow under the diaphragm.

Δ - NEC

↳ In 90% preterm

2nd - 3rd wk of life.

Q. Features of NEC are all except

A) Abd. distention

B) ↑ Bowel sound

C) Pneumoperitoneum

D) Metabolic acidosis

Rx: NEC

- Stop all oral feeds
- TPN (Glucose, AA's, Lipids)
- Antibiotics (Cefotaxime; Vancomycin; Metronidazole)
- Stage III - may require Sx.



Q. Child w/ NEC w/ perforation & poor general cond' is treated w/ :

- A) Conservative t/t only.
- B) Frank drain w/ glove
- C) Laparotomy w/ resection anastomosis
- D) Extracorporeal membr oxygenation.

Stage III
 Stable - Laparotomy
 Unstable - Peritoneal drain

NEONATAL SEPSIS:

= Symptoms + Bacteremia

EARLY

< 72 hrs

LATE

> 72 hrs.

R/F - Maternal Fever < 7 days

M/c/c - Nosocomial

Foul liquor

M/c/c world - Coagulase -ve staph

PPROM: Chorioamnionitis.

In India - Klebsiella, S. aureus.

M/c/c world: Group B streptococcus, E. coli
(M/c)

Meningitis \Rightarrow CSF exam

Rx - Ampicillin + Gentamycin

- Cefotaxime + Amikacin

M/c/c India: Klebsiella, S. aureus

Sepsis screen for early diagnosis:

① TLC < 5000/cumm or > 20000

② ANC < 1500/cumm

③ PS for band cells / Immature neutrophile > 20% ($I/T > 0.20$)
& toxic granules

④ Micro ESR ($B_{min}^{(N)} - 3$ days of life) > 15mm fall in 1st hour

⑤ CRP : Procalcitonin +ve

⑥ Lumbar puncture (In late sepsis)



(7) Chest X-Ray

ANC = Neutrophil + Band cells

Q Lab finding in Neonatal Sepsis except -

- A) ↑ CRP
- B) Leucocytosis
- C) ↓ ESR
- D) Toxic granulated multilobulated nuclei

Duraction of Antibiotics in Neonatal Sepsis:

Bacteremia = 10-14 days

Meningitis = 21 days

Arthritis, Osseous myelitis = 4-6 wks

Temp regulation of newborn:

Non-shivering Thermogenesis -

Brown fat

- Nape of Neck
- Interscapular
- Around kidneys & adrenal
- Around blood vessels (around mesentery).

Axillary temp:

- Normal $36.5 - 37.5^{\circ}\text{C}$
- Cold stress / mild hypothermia ($36-36.4^{\circ}\text{C}$)
- Moderate hypothermia ($32-35.9^{\circ}\text{C}$)
- Severe ($< 32^{\circ}\text{C}$)
- Hyperthermia $> 37.5^{\circ}\text{C}$ - mostly iatrogenic.

Prevention of hypothermic: KMC (Kangaroo mother care).



Q. All components are of KMC except:

- A) Kangaroo position
- B) Kangaroo nutrition
- C) Early discharge & follow up.
- D) Supplementary nutrition.

Q. When to stop KMC:

- When a child reaches 2500 gm wt.

Q. Delivery room temp - $> 72^{\circ}\text{F}$ (AAP)

$> 25^{\circ}\text{C}$ (WHO)

$\hookrightarrow (25-28^{\circ}\text{C})$

AIIMS NICU protocols

- $< 28\text{ weeks} / < 1000\text{ gms} \rightarrow$ put baby in Polythene bag.

reduces convection

- Incubator \rightarrow Reduces convection.

Q Mechanism of heat transfer in overhead radiant warmers :-

Radiation, + ↓ convection loss.

Hypothermia:

CNS : Asphyxia

CVS : Asphyxia Cardiomyopathy

Lungs : ARDS

Kidneys : Asphyxia ATN

Hypoglycemia ; HypoCa ; HypoMg .

Retrobulbar Fibroplasia /

ROP (Retina of Prematurity) :

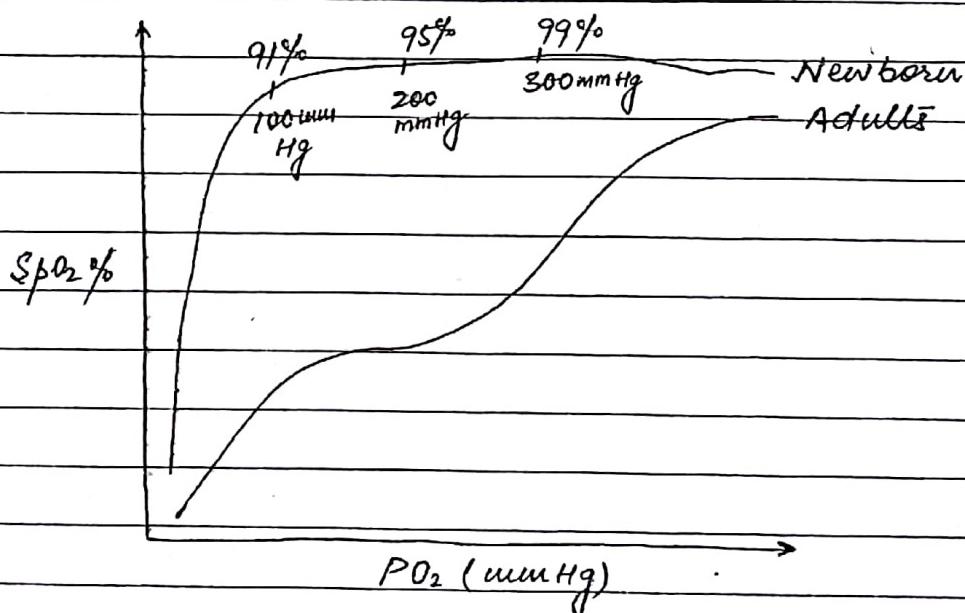
R/F : Preterm.

Higher flow O₂.

- Proliferation, dilatation & tortuous vessels.
- Tractional retinal detachment.
- ROP stage I-V : 'plus' - Blindness.

Stage IV : Incomplete tractional RD

Stage V : Complete " "



WHO targets SpO₂ = 91-95% in preterm.

(Inv. of ROP)

BLINDNESS : Regular indirect ophthalmoscopy.

AAP ROP guidelines - (Risk < 30 wks / < 1500gms)

Gestation(wks) 1st visit to ophthalmologist

Post menstrual(wks) Interval (wks)

22

31

9

23

31

8

24

31

7



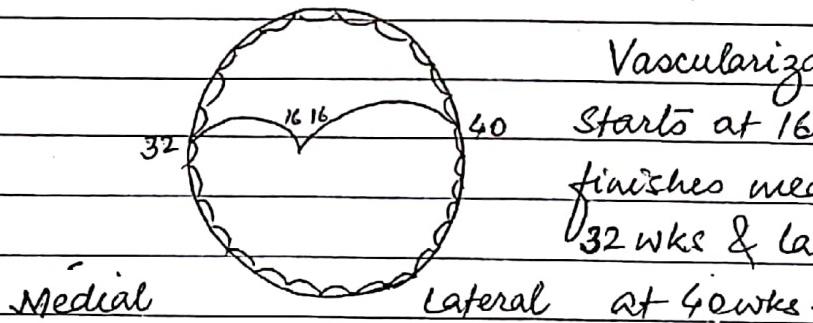
25	31	6
26	31	5
27	31	4
(28)	32	4
29	33	4
30	34.	4

Q. Pediatrician in a district hospital calls ophthalmologist for:

- a) Newborn c Respiratory distress
- b) Newborn 28 wks gestation

↳ After first visit, baby has to go to ophthalmologist, every 2wks till his/her eye look like term retina.

Retinal Vascularization: Left eye.



Rx: ROP

- Laser Photo coagulation / Peripheral ablation
- Type I ROP (all plus disease)
- Cryotherapy.
- Stage V → Retinal reattachment.

New drug - Bevacizumab → Anti-VEGF ; resistant.

Respiration :

Tachypnea : RR > 60/min.

Silverman Anderson Retraction Score :

Feature	Score 0	1	2
Chest movement	Equal	Respiratory lag (upper chest inspiration)	Paradoxical/ Saw Respiratory
Intercostal Retraction	None	Minimal	Marked
Xiphoid Retraction	None	"	"
Nasal Flaring	None	"	"
Expiratory Grunt.	None	Audible c Stethoscope	Audible

Downes scoring for Respiratory distress :

Feature	Score 0	1	2
RR (per min)	< 60	60-80	> 80/ apneic episode
Cyanosis (central)	None	In room air	in 40% O ₂
Retraction	"	Mild	Moderate - Severe
Grunting	"	Audible c Stethoscope	Audible c Stethoscope
Air entry (mid-axillary line)	Clear	↓ (Delayed)	Barely audible



M/c/c of RD → Surfactant deficiency

Surfactant → Phosphatidyl choline (65%) (Most imp.)

Phosphatidyl glycerol

Phosphatidyl inositol

Phosphatidyl ethanolamine

SP-A, SP-B, SP-C, SP-D.

Other protein.

↳ Homogenates - 20 wks gestation

Ammiotic fluid - 28 wk gestation

Mature levels - 95% 35 wks.

RDS risk OC Degree of prematurity.

Risk of RDS:

< 28 wks - 60-80%

> 37 wks - < 5%

Infant of diabetic mothers, ^{at} term

Rare in IUGRs → Stress → Cortisol

Lechithin: Sphingomyelin ratio

> 2 → lung mature.

> 3.5 infants of DM mothers.

CXR: RDS (lungs) : Ground Glass appearance.
(white out lung).

Air bronchogram - Classic feature of RDS.

(N) CXR seen in Early RDS.

Rx : • Humidified O_2 (40-60%)
 - We don't give 100% O_2 bcoz of ROP.

- Nasal CPAP : mild - moderate distress
 early in ELBW's

F_iO_2 : Start at 40-60% -----> room air

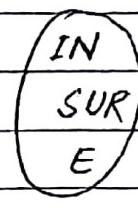
PEEP : 5 cm H_2O

- Intratracheal surfactant - Severe ; as rescue.

- Intubate the baby

Give Surfactant

Extubate the baby



Survanta - Bovine

Curosurf - Porcine

Infasurf - Calf.

Synthetic also available

Q

32wks, Preterm baby in emergency C.S.

Grunting, RR = 70/min. Best management of choice

A) Humidified O_2 by hood

B) Mechanical ventilation.

c) CPAP

~~✓~~ Surfactant therapy & mechanical ventilation.

Q

All occurs in RDS except:

A) Cyanosis

B) Occurs in preterm

C) More in IDM

~~✓~~ Treated by 100% O_2



Q. All true about CPAP except :

- A) Initiated $\text{FiO}_2 0.40 - 0.60$
 - B) Used in apnea of prematurity.
 - C) Improves compliance.
 - D) Volume ; $\text{FRC} \uparrow [\text{RDS FRC} < \text{CV}]$
 - E) Used prophylactically in ELBW's.
- All true.

Q. Term female, Birth wt. = 3.5kg, uncomplicated delivery.

Respiratory distress after birth.

CxR - Ground glass appearance.

On ventilation & given surfactant.

But cond'n deteriorates & hypoxemia increases.

H/o sibling dying c in one week c similar complain. ECG & blood culture - N.

↳ Δ = Neonatal Pulm. Alveolar proteinosis

↳ Autosomal Recessive

↳ Mutation in Protein B (Rarely in c)

Rx : Early lung transplantation

Postmortem Rx - Pink eosinophilic material in lung.

- Idiopathic : 90%

Adult or acquired

IgG antibodies to GM-CSF

- Secondary - 5-10%

Hematological malignancy

Inhalational lungds.

Silicon

Titanium Oxide.

- Congenital : 2%

CT scan - crazy paving pattern
(Prominent intraalveolar septae).

Rx - Early lung transplant in neonates.

Adults → Broncho alveolar lavage.

Respiratory distress : Newborn

Tracheo-esophageal fistula (TEF)

M/c type - Type C

Distal TEF



Esophageal atresia

- Cyanotic newborn & frothing
- Aspirates gastric juice lead to pneumonia.
- Not a surgical emergency.

Rx - Keep him propped up.

- Suction catheter in upper blind esophageal pouch.

Diaphragmatic hernia:

85% cases - left.

pushed heart & trachea - to opposite side.

- Scaphoid Ab domen
- Barrel chest
- Mediastinal shift to right.
- Apparent dextrocardia
- Peristalsis on left chest.

Diaphragm - develops from septum transversum
& pteroperitoneal canals.

↳ fail to close on left side

(Bochdalek hernia)

- < 5% are B/L



Q. Cause of death in Congenital Diaphragmatic Hernia:

A) Septicemia

B) Pulmonary hypoplasia - Left

C) Hemorrhage.

Bag & mask is absolutely c/i bcoz it will cause further abdominal distension.

↓
So elective intubation is done.

Baby born → Bag & mask & 100% O₂

↓

diaphragmatic hernia

↓

Intubated

Next step - NG tube to decompress the gut.

Born baby → Diaphragmatic hernia

↓

Intubated

↓

Heart further to right

Next step - Remove the tube
& Reintubate.

Q. Most imp. Prognostic factor in Congenital diaphragmatic hernia:

A) Pulm HTN - Persistent (PPHN)

B) Age

C) Time of Sx

D) Size of defect.



Prevention : PPHN

- Elective intubation
- HFOV $> 300 - 600 / \text{min}$

Rx : PPHN

- iNO \rightarrow Pulm. vasodilator.
- Sildenafil PDE-V inhibitor

New drug : Bosentan, Ambrisentan — Endothelin antagonist.

- $\text{PGI}_2 \Rightarrow$ Iloprost
- Amlodipine ; ECMO (Extracorporeal membr oxygenation)

Transient Tachypnoea of Newborn (TTNB) :

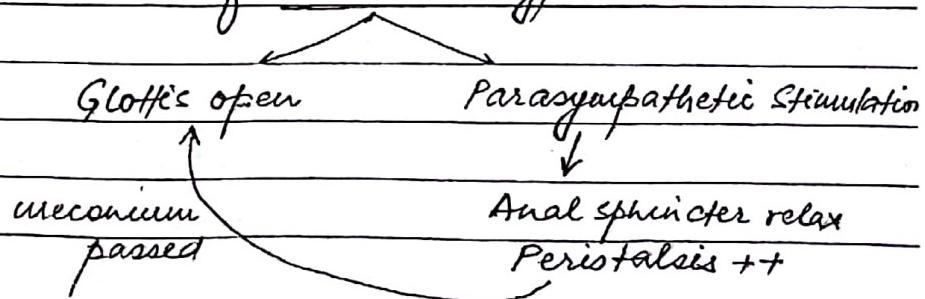
- R/F : • Term by C.S. (lungs are wet)
• Macrosomia
• Excessive maternal sedation.
• Precipitous labour.

CxR - a prominent horizontal fissure
↳ Most specific feature.

- Benign cond'n
- Self limited : 48-72 hrs.
- FiO_2 requirement < 0.40
- Never require mechanical ventilator.

Meconium Stained liquor :

Meconium is marker of Perinatal Hypoxia

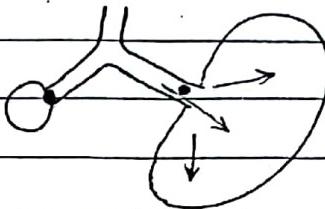




Perinatal hypoxia : Common ein posterum
scuz of VPI.

Meconium :

Physical -



- ball valve mechanism
- air leak 20-30%

Chemical - Irritant → pneumonia

↳ Impair surfactant funcⁿ.

Biological - Good culture media

Meconium Stained liquor.



Baby born

Vigorous

Tone is good

Respiratory effort is good

HR > 100/min

yes

No

Transfer

the baby to
mother

Catheter

in the nose



PPV @ 100% O₂.

Q Sequence of Resuscitation

A) Mouth → Nose

B) Nose → Mouth

C) Mouth → Nose → Trachea

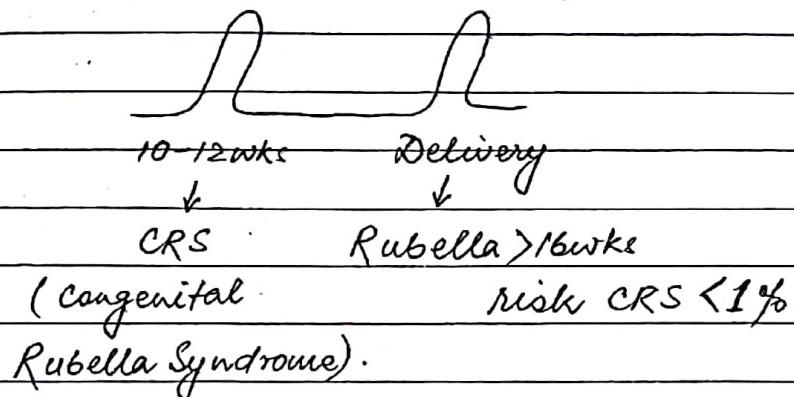
Intrauterine TORCH infections:

Others - HIV, HBV, Varicella, Syphilis.

Common features:

① - Asymptomatic

Rubella: Transmission - 2 peaks



② - If symptomatic

- SGA; failure to thrive
- Anaemia; thrombocytopenia
- Hepatosplenomegaly
- Unexplained Rash & cholestasis.

△ - IgM (evidence of infection)

IgG persisting beyond 6-9 months

HIV in infants

- Infants can't make IgM-HIV or IgA-HIV.
- Maternal IgG-HIV can persist in the baby for 18 months.
- HIV < 18 months : Diagnosis.

Best : DNA qPCR

P24 assay

Culture is difficult.



Intracranial calcification & Chorioretinitis] M/c/c - Toxoplasmosis - CMV

Toxoplasmosis

- 25-50%
- Choroid plexus calcification.
- Subependyma & Cadate nucleus calcification.
- ~~Hydrocephalus~~ Hydrocephalus
- Seizures.

CMV

- Periventricular Calcification.
- Atrophy of brain (Cortical).
- Hydrocephalus ex vacuo.
- M/c/c of Non syndromic SNHL
- Seizures
- Microcephaly
- Mental Retardation.

Q. Pregnant lady; no complain. Mild cervical lymphadenopathy in 1st trimester. Prescribe

~~(Spiramycin)~~ but she was non compliant. Baby born

prevent vertical transmission of hydrocephalus & intracerebral calcification.

↳ Toxoplasmosis

DOC: Rx: Pyrimethamine + Sulfadiazine.

△: Best: IgM Immunosorbent assay.

Sensitivity of ELISA:

IgA >> IgM

Q. True about transplacental

CMV infection:

- It is M/c/c of non-syndromic SNHL.

Q. Doesn't establish Δ of CMV in neonate -

A) Urine culture of CMV

B) IgG CMV antibodies in blood

C) Intra-nuclear inclusion

bodies in hepatocytes (Owl-eye)

D) CMV viral DNA in blood

by polymerase chain reacⁿ.

Best specimen - Urine culture

& Saliva.

CMV disease: Retinitis, colitis, pneumonia.

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Rx : CMV

DOC : i.v. Ganciclovir (Severe, child, pregnant)

- Oral; Prodrug → Valganciclovir
- Resistant to oral Foscarnet.

Congenital Syphilis:

EARLY

- C in 1st 2 yrs of life.
- Mucocutaneous rash/rhininitis (Snuffles).
- Lymphadenopathy.
- Hematological (Autoimmune Anemia)

LATE

- After 1st 2 yrs.

Hutchinson's Triad:

- ① Hutchinson teeth/
mulberry molar - 1st
lower molar.
- Saddle nose, Frontal bossing,
Olympian's brow,
- ② Higoumenaki's Sign
(Sternoclavicular prominence)
- Rhagades
- ③ Nerve deafness. (SNHL)
- Cluttons joints
(painless joints)
↳ Risk of injury.

Pseudoparalysis:

M/c/c - Scurvy

- Early syphilis
- Osteosyphilitic
- Septic arthritis.
- Hypokalemia → Hypotonia.

DOC - Penicillin G

→ 10-14 days



Rubella Syndrome :

Trisomy — Microcephaly (Mental Retardation)

PDA

Cataracts.

M/o eye manifestation of Rubella — Salt & pepper fundus

Cataract

Glaucoma

Micro-ophthalmia

HEART — PDA

Peripheral pulm. stenosis

VSD

ASD (Rare).

Q. Rubella embryopathy except.

A) Deafness

B) MR

~~C) AS~~

D) PDA

Q. True about Rubella embryopathy except:

A) Diagnosed when IgM antibodies in child.

~~B) Infection after 16 wks results in major congenital anomalies.~~

C) Deafness, heart disease, Cataract.

Q. Hypoplastic limb — Varicella (Chicken pox) embryopathy during pregnancy.

Varicella Embryopathy -

- Skin Rash
- Optic nerve hypoplasia
- Brain - Cortical atrophy.
- LS plexus - Aplasia / hypoplasia limbs.

Mother get chick pox - 5 days before delivery
or, \leq 2 days of delivery.



Baby chicken pox illness.



Prevention : Varicella Zoster Ig to the baby.



even after 120 hrs of exposure.

Pregnant ; HBsAg +ve : no jaundice

HBeAg +ve \Rightarrow 90% chance baby is carrier

& later in life Portal hypertension.



Ascites, Splenomegaly, Varices

HBeAg -ve \Rightarrow Anti HBeAg +ve

then 10% chance of Vertical transmission

Give HBIG baby to 12 hrs of life

HBV vaccine to baby \leq 24 hrs of life.

Pricked by HBsAg +ve patient

Are you immunized?

~~Anti-HBc IgG~~ - Anti HBs Ab titre

Good $> 10 \text{ mIU/mL}$; High risk > 100



Incomplete vaccination / Not know titre



HBV + HB Ig
a/c to CDC guidelines.

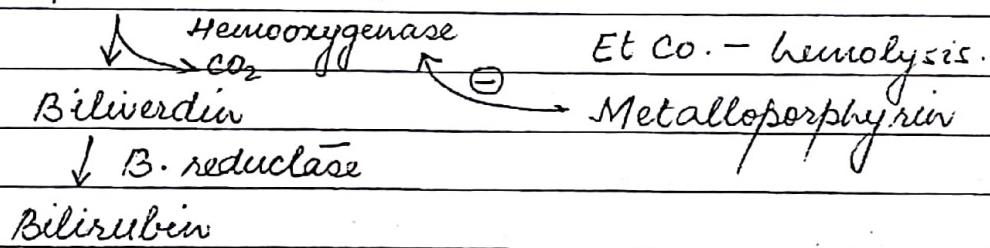
If titre is good - don't do anything.

CDC HBV DNA load $> 1000 \text{ IU}/\text{ml}$

↳ Can't gain Surgical branch.

Neonatal jaundice

Mean:



Bilirubin

1 gm of Hb = 34 mg of bilirubin

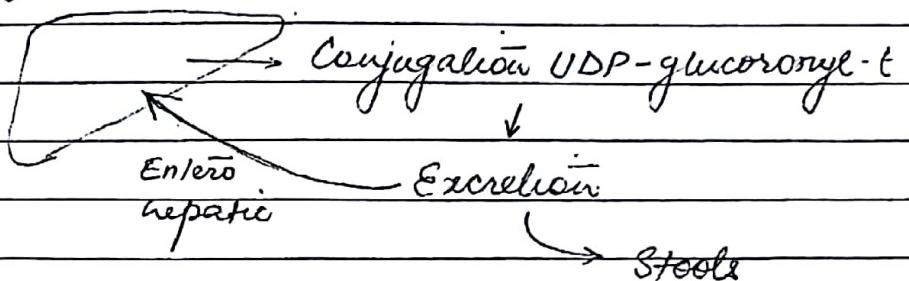
1 gm/dl Albumin binds to 8 mg bilirubin

- Unconjugate bilirubin passes BBB & cause jaundice & Kernicterus.
- In 1st 2wks, BBB is not developed properly.
- (N) S. Albumin = 3.5 - 5.5 g/dl
- Healthy term baby can bind 24 - 25 mg/dl bilirubin.
- Sick, preterm, risk factors can go into early Kernicterus.
- In adults ammonia crosses BBB in hepatic encephalopathy.
- Healthy Baby ($< 1000 \text{ g}$) \rightarrow We start phototherapy ($5-7 \text{ mg/dl}$)
↓
Bilirubin
- Sick baby \rightarrow Bilirubin ($4-6 \text{ mg/dl}$) - we start phototherapy.

Bilirubin



uptake by liver (γ -ligandin uptake)



$> 2 - 2.5 \text{ mg/dl}$ in adults \Rightarrow Yellow Sclera.

Krausser's zone:

progression of jaundice in new born is
cephalo-caudal
(Bilirubin)

Zone I (5 mg/dl) \rightarrow

Zone II (10 mg/dl) \rightarrow

Zone III (12 mg/dl) \rightarrow

Zone IV (15 mg/dl) \rightarrow

Zone V ($> 15 \text{ mg/dl}$) \rightarrow

↳ Danger Zone.

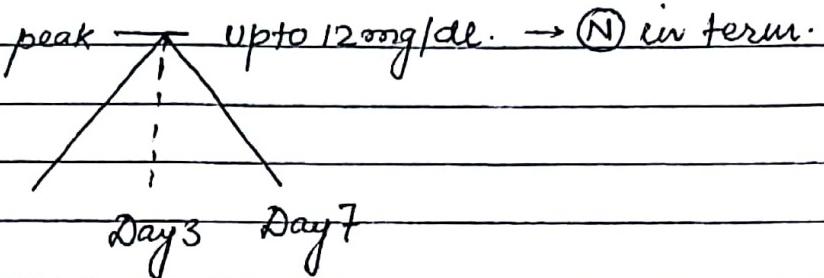
6 causes of physiological jaundice:

- ① Life span of RBC is less.
- ② Hematocrit more
- ③ Newborn deficient in γ -ligandin.
- ④ UDPglucuronyl-t deficient in newborn.
- ⑤ Excretion reduced.
- ⑥ Enteric hepatic circulation ↑.

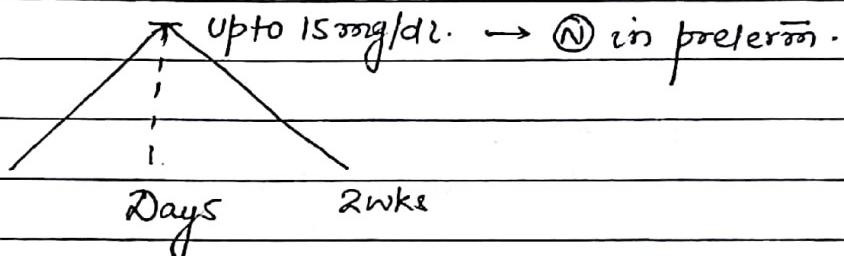


Physiological jaundice:

jaundice in term babies.



Preterm - have more jaundice.



Pathological jaundice:

- Hemolysis.

- M/c/c - Incompatibility Rh/ABO [O mother; baby A/B]
- RBC ~~membrane~~ membrane defect.
- RBC enzyme defect.

Def: jaundice c in 24 hrs of life.

* Rh-ve mother; previous abortion we take cord blood samples.

- Rh status of baby. → +ve
- Hb → 10 mg/dl
- Bilirubin → 5 mg/dl
- Peripheral Smear (P/S) +ve
- Direct Coombs test (DCT) → +ve



means Severe hemolysis

Rx → Exchange transfusion at birth.

① jaundice in 24 hrs of life.

② Reaching 20mg/dl

③ Rate rise > 0.2mg/dl/hr.

④ Persisting beyond

- 1 wk term

- 2-3 wks preterm

⑤ Clay stools.

Phototherapy:

- Any jaundice on day 1 of life start phototherapy -
Serum bilirubin cut off.

Phototherapy

Exchange transfusion

Healthy babies / Babies < R/F Healthy babies / Babies < R/F

	← Any visible jaundice →	260(15)	220(10)
Day 1	260(15)	170(10)	425(25)
Day 2	310(18)	250(15)	425(25)
Days 3			340(20)

R/F ① Gestation < 35 wks / wt. < 2kg.

② sepsis

③ Hemolysis

④ Asphyxia

⑤ Sick baby.

Principle:

- Structural isomerization

• Bilirubin → LUMIRUBIN

- Photo isomerization

• $4Z15Z \leftrightarrow 4Z15E$ (soluble).

- Minor pathway → photo-oxidation.

• 40 cms away, falls @ 4-6 mg/dl/day.



- Phototherapy occurs at 425-475 nm of blue green light.
- Irradiance \rightarrow 6 micro watt/cm²/nm
- Intensive $>$ 30 micro watt/cm²/nm.

Q

AIIMS Nov. 2013

Which does not effect the efficacy of phototherapy?

- A) Types of phototherapy lamp.
- B) Skin pigmentation.
- C) Spectral radiance of incident light
- D) Initial bilirubin levels.

#

Complication:

- Hyperthermia; insensible losses
- Hypocalcemia
- Diarrhoea.
- Cover eyes & genitals
 - Retinal damage
 - Mutations.
- Phototherapy is c/i in conjugated jaundice

↓
Bronze baby syndrome.
(Skin, Urine).

Exchange Transfusion:

- Double volume exchange transfusion.
- # Blood vol. of new born = 80 mL/kg
- $\rightarrow 2 \times 80 \text{ mL/kg} = 160 \text{ mL/kg}$
- Transfuse fresh (< 7 days) whole blood.
- Reduces bilirubin by 85%.

↓
Not 100% because bilirubin is also in tissue.

- Albumin ↑ efficiency.

Complications:

- Infection
- ACD (Acid citrate dextrose)



Bicarbonates

1 mole of citrate → 3 mole of Bicarbonate.

- Metabolic alkalosis
- Hypokalemia, hypocalcemia.
- Old blood → Hyperkalemia, metabolic acidosis.

Persistent jaundice:

Cause - Hypothyrodesim

Breast milk jaundice

Hematoma (Cephalhematoma, IVH)

Clay colour stool (cholestasis)

Cripple - Naggar syndrome type II (milder form)
 ↳ deficiency of UDP-glucuronyl-

Cripple - Naggar Syndrome type I - Very severe

↳ Absent UDP-glucuronyl-t.

Pathological jaundice

Breast milk jaundice

- Onset → day 14
- Some mother have pigment
- ↳ inhibit conjugation.
- Day 14 = 20-30 mg/dl
- May Kernicterus.
- Persists 4-6 wks.

Breast feeding jaundice

- Onset → Day 3.
- In primigravida.
- Starvation stimulates enterohepatic circulation.



Rx: Temporary interrupt
48-72 hrs.

Rx: Ensure feeding.

Meanwhile give
formula milk.

Q True about jaundice in newborn (neonates) is -

- A) Can be seen after Ventouse delivery.
- B) Physiological jaundice seen c in 48 hrs of birth.
- C) Increased conjugated bilirubin leads to kernicterus.
- D) Breast milk jaundice is maxⁱⁿ in 7 days of birth.

Neonatal cholestasis:

Neonate, jaundice & clay stools.

- Direct bilirubin $> 2 \text{ mg/dl}$ or $> 20\%$ total bilirubin

Medical - Common.

Neonatal hepatitis → CMV

Sepsis

Galactosemia

α -1 antitrypsin deficiency.

Neonatal hemochromatosis.

Surgical - Extra hepatic

biliary atresia (EHBA)

Rx - Kasai's surgery c in
8wks of life otherwise

80% die.

↓

M/c/c of indication of
liver transplant in babies
→ EHBA.

Q Which is an ominous sign in a 10 day old newborn?

- A) Unconjugated hyperbilirubinemia
- B) Conjugated "
- C) Failure to gain wt.
- D) Doll's eye reflex
↳ Normal in 1st 10 days of life.

GGT → Gamma Gutaaryl transferase.

D312 1 47
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Medical - Common

Surgical - EHBA

- GGT ten times higher in Surgical causes.
- Do USG → Shows Intra hepatic biliary radicle (IHBR) are dilated.
- Triangular cord sign 

- HIDA nuclear Scan:

HIDA dye not seen in gut, even in delayed images; while in hepatitis excretion of dye.

Best test - Liver biopsy.



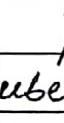
Show dilation & proliferation of intrahepatic bile duct.

Before doing HIDA Scan we have to give phenobarbitone 2 to 3 days before test.

Gold Standard - Preoperative Cholangiography.

Alagille Syndrome -

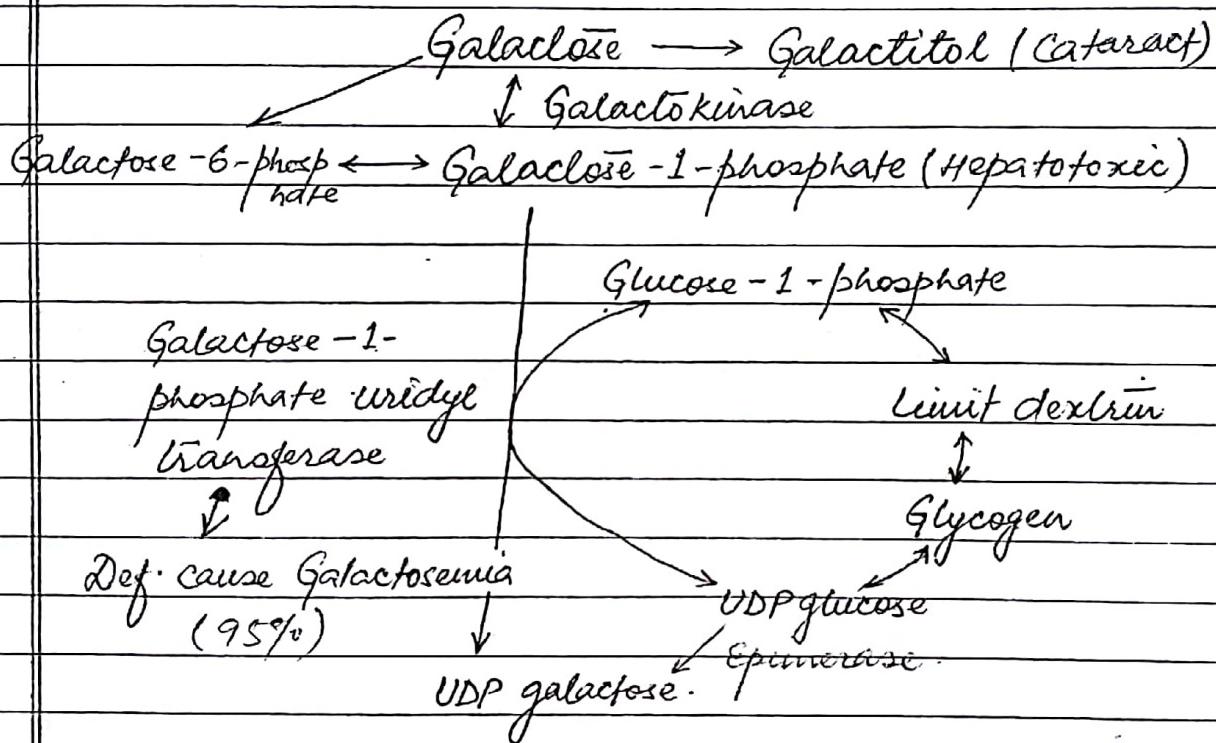
- Cholestasis
- AD
- Bile duct paucity Syndrome.
- M/c heart disease → Peripheral pulm. stenosis.

Rubella 



- Q. 1 month old child present with conjugated bilirubinemia & intrahepatic cholestasis. On liver biopsy staining with PAS, red coloured granules were seen inside the hepatocytes. Probable diagnosis is -
- A) D-1-antitrypsin deficiency.
 B) Congenital hepatic fibrosis
 C) Hemosiderosis
 D) Wilson's ds.

CMV - Intracellular "owl eye" inclusion bodies.



- He get jaundice, bleeding, hypoglycaemia, PTT.
- Galactitol cause cataract.
- Rx - lactose free milk.
- Galactokinase deficiency → Only cataract
 No liver failure
- Fructokinase def. → Causes Benign glycosuria
 ↳ No symptom.

Frucone-1-phosphate leads to liver failure.

Date 1/1
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Q. M/C/C of Neonatal Cholestasis:

- A) EHBA
- B) Neonatal hepatitis
- C) Choledochal cyst
- D) Physiological

Q. Neonatal cholestasis seen in -

- A) Chronic hepatitis (>6 month)
- B) Hep. B
- C) Galactosemia
- D) Rh incompatibility (cause unconjugated jaundice)

Q. Pregnant lady, HBsAg +ve, No jaundice.



next step \rightarrow HBeAg

- HBeAg +ve (90%)

Carrier HBsAg \rightarrow Portal HTN



Triad - Ascites

Splenomegaly
Varices.

- HBeAg -ve

anti HBe Ab \oplus

\hookrightarrow 10%

Immunization [HB Ig - baby c in 12 hrs of life

\downarrow HBV - baby in 24 hrs of life

prevent Vertical transmission



CDC guidelines:

Pricked by HBsAg + patient



Are you immunized?



Anti-HBs Ab titre

Good $> 10 \text{ mIU/ml}$

High risk $> 100 \text{ mIU/ml}$

Don't know titre \rightarrow Incomplete



HBV + HB Ig

If Good titre \rightarrow Nothing is to be done

Q.

New born, Respiratory distress:

Neonatal Seizures:



Neonatal hypoglycemia:

Causes:

- limited stores → Preterm, IUGR's
- Stress → Sepsis
- Polyhydramnios
- Galactosemia - liver failure.
- Low cortisol - CAH (Congenital Adrenal Hyperplasia)
- Hypopituitarism
- low ACTH; low LH/FSH → Micropenis.
- Hyperinsulinism - In infants c diabetic mother.
 - ↳ Foetus c β-cell hyperplasia.

Foetus Blood glucose ↓
↑

Pederson's hypothesis

- Insulinoma; Nescidioblastosis
- Beckwith Wiedemann syndrome.
 - ↳ Hemi hypertrophy of limb
 - ↳ Macroglossia
 - ↳ Risk of Wilms
 - ↳ Hyperinsulinism

Q. A term baby to a diabetic mother, few hours after birth - was lethargic & his blood glucose was 30 mg%. What should be done next -

A) Give 10% dextrose orally.

B) 10% dextrose i.v. - Bolus 2ml/kg → Glucose drip

C) Give expressed breast milk

D) DO exchange transfusion.

GIR
↓
6-8 mg/kg/min

Neonatal hypoglycemia:

Symptomatic

→ Bolus 2ml/kg 10% dextrose

if by Glucose drip

6-8 mg/kg/min

Asymptomatic

→ Blood glucose < 20 mg/kg

- ↓

Glucose drip (6-8 mg/kg/min)

→ Blood glucose = 20-45 mg/dl

↓

Breast feed; 1 hourly.

A baby on glucose drip & he gets seizures

how to ↑ glucose rate

- Up to 12.5% through peripheral vein.

Maxth glucose infusion rate (GIR) = 12 mg/kg/min.

Emergency drug = i.m. Glucagon.
for hypoglycemia

↓
Glycogenolysis
+ Gluconeogenesis.



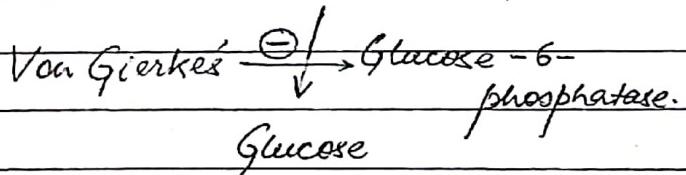
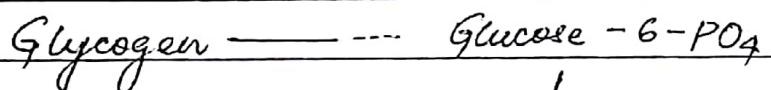
Q. Glucagon is effective for Mx in persistent hypoglycemia in all except.

- A) Large date for baby.
- B) Neuridioblastosis
- C) Galactosemia
- D) Infant of diabetic mother.

Q. 1 yr old, hypoglycemia & hepatomegaly
No jaundice.

Hypoglycemia doesn't respond to glucagon.
↓

Δ → Von Gierke's (Glycogen storage ds - I)



Types of Glycogen storage disorder:

V = Von Gierke's → Liver primary

P = Pompe's ds → Heart primary; Cardiomegaly.

C = Cori → Debranching enzyme. Large QRS complexes.

A = Anderson → Branching " deficiency

M = Mc Ardles → Muscle phosphorylase deficiency.

Harding =

Ton =

Jaundice never occurs in Glycogen storage ds.

Rx Pompe's: Enzyme replacement therapy.

Enzyme absent in Pompe's → lysosomal α -1,4-glucosidase
— Also called Acid & neutral maltase.

Muscle affected in GSD:

Calcium:

(N) S. Ca^{2+} → 9-11 mg/dl.

Neonatal hypocalcemia:

- S. Ca^{2+} < 7 mg/dl
- Best index of body calcium

Ionized < 4 mg/dl (or) < 1 mmol/L

- Tetany is rare in infants.

- Tremors, seizures, jitteriness.

↳ Tremulousness is stimulus sensitive.

↳ Can stop on passive restrain.

Early Hypo Ca^{2+}

Causes - Prematurity
Asphyxia

↳ Infant of DM mother.
↳ Test:

Blood glucose

+ S. Ca^{2+} & S. Mg^{2+}

Late Hypo Ca^{2+}

Cause - Feeding c phosphate rich milk (cow milk)

Good Ca^{2+} Supplement ($\text{Ca}^{2+}/\text{P} > 2$)

100 ml

Ca

P04

Cow milk

118 mg

100 mg

Breast milk

34 mg

15 mg

Advantages of Breast milk:

- protects against late onset hypocalcemia.
- protect against pneumonia
- NEC (Necrotizing Enterocolitis).
- Allergy, Eczema, asthma.
- Rota virus diarrhoea.
- Bronchiolitis (IgA-RSV)

Q. Milk deficient in :

- a) Iron & Vit. C

Q. Breast milk has enough iron & Vit. C for 6 months.

∴ Scurvy never occurs in ^{1st} 6 month of life.

Q. If baby on exclusive breastfeed for 2 yrs.

Iron deficiency anaemia.

Q. APP recommends Vit. D to all infants - Vit. D drops.

- RDA Vit.D infant 400 IU/day
- Breast milk 25 IU/L.

M/c/c of HypoCa²⁺ in infants = Maternal deficiency of Vit.D.

25[OH] Vit.D = Status.

Infant of DM mother:

- Can be stillborn; preterm
- Macrosomia
 - ↳ Bcoz of Hyperinsulinism
- Linear growth in utero depend upon insulin & or Insulin like growth factor.
- IUGR → White's classification class F/R
mother → Placental vasculopathy.
- Hypoglycemia
- Hypo Ca^{2+} , Hypo Mg^{2+}
- Neonatal jaundice
- Polycythemia → Renal Vein Thrombosis (RVT)

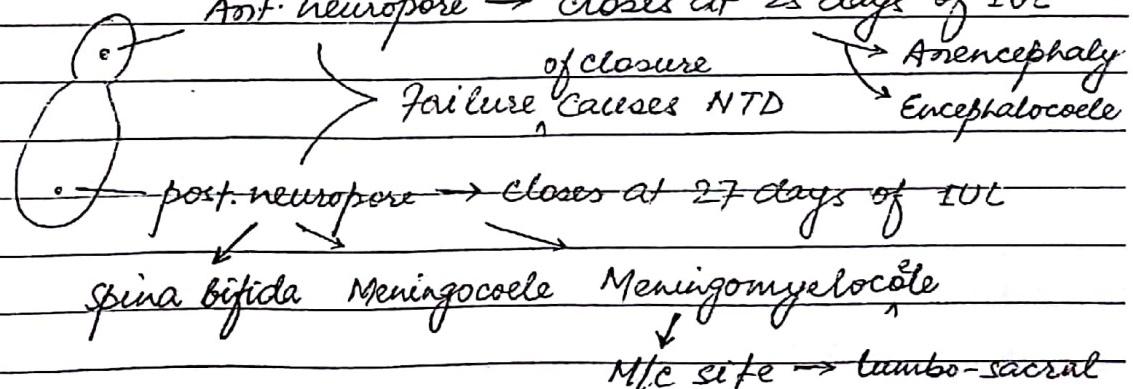
Q. Atq. Not seen in infant of diabetic mother:

\downarrow
Hyperglycemia.

Anomaly:

- (M/c)- CVS - 8.5% [VSD; HOCM] Asymmetrical septal hypertrophy
- Neural tube defect \rightarrow 5% hypertrophy
 - Lazy left colon syndrome \rightarrow Pseudoobstruction of colon
 - Sacral agenesis/Caudal regression Syndrome
 - ↳ Most specific.

NEURAL TUBE DEFECT (NTD)





Anecephaly:

- Ant. neuropore fails to close.
- No brain, absent of part of hindbrain.
- Earliest abnormality diagnosed by USG
(10-12 wks)
of gestation.
- Most severe NTD
- Don't resuscitate
- Mostly post-term.

Herniation of brain tissue — Encephalocele.

Lumbo-sacral myelomeningocele:

Complication:

- Paraplegia/Paraparesis
- Neurogenic bladder → CKD.
- Constipation (Severe)
- Associated hydrocephalus. (obstructed)

Bcoz of Arnold-Chiari type II malformation

↓
MRI Brain

Ruptured myelomeningocele:

Best test — Blood culture.

- Cover c Normal saline soaked gauze.
- 95% of neonatal meningitis have leukemia.
- M/c/c of meningocele — Folic Acid deficiency.

Folic acid deficiency:

- 1/ + 3 months conception - start folic acid.
- 400 mcg given.

Recurrence:

1 child - 3.5% chance

2 child - 10%

3 child - 25%

To prevent recurrence → Folic acid 4mg

↓
reduces risk by 75%.

△ : ① USG

② Amniocentesis

↳ Acetylcholinesterase

& α -fetoprotein are markers.

α -fetoprotein in mother serum is marker of Neural tube defect.

Resuscitation:

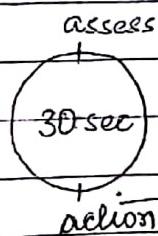
NRP Guidelines 2015:

T = Temp

A = Airway : position neck, suction

B = Breathing

C



Indication of Bag & mask \bar{c} 100% O₂:

- Apnea/gasping ~~after~~ after initial steps.
- HR < 100/min after 30 sec PPV.
- Central cyanosis despite 100% O₂.
- Chest compression if HR < 60/min, falling after 30 sec PPV.
- Chest compression : Bag mask = 3:1

In 1 minute = 120 events

90 Chest compression & 30 Bag & mask.

Compression to Ventilation ratio:

- Children/Infant - Single rescuer 30:2
2 rescuer 15:2
- Adults - 1 or 2 rescuer 30:2

CPR sequence \rightarrow CAB

Drugs for resuscitation:

- 1) 0.9% NaCl 20ml/kg bolus \rightarrow shock
- 2) 1:10,000 epinephrine 0.1-0.2ml/kg

↓

If HR = 0 or falling.

- 3) I.V. NaHCO₃ - documented metabolic acidosis.
- 4) I.V. Naloxone - mother opioid addvt.

Targeted preductal SpO₂ after birth:

1 min = 60% - 65%

2 " = 65 - 70 %

3 " = 70 - 75 %

4 min = 75 - 80%

5 min = 80 - 85%

10 min = 85 - 90%

GENETICS

  → Heterozygotes for AR carrier

 → Carrier for sex-linked recessive

 → Death

 → Abortion or stillbirth
Sex unspecified.

 → Proband.

  → Consanguineous marriage

  → Dizygotic twin

  → Monozygotic twin

  → azoospermia

 → Endometriosis

  → Infertility.

   → Adopted in

   → Adopted in

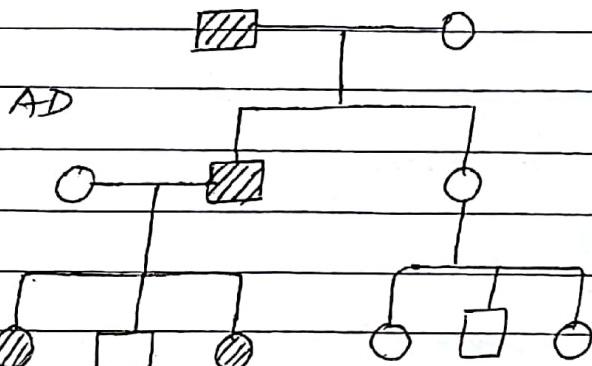


→ No children for choice
or reason unknown

→ Vasectomy

→ Tubal

→ Divorce.



Autosomal dominant:

D = Dystrophy myotonie → distal myopathy.

O = Osteogenesis imperfecta

M = Marfan Syndrome

I = Intermittent porphyria

N = Noonan Syndrome → Turner phenotype XX & XY.

A = Adult PKD, Achondroplasia

N = NF (Neurofibromatosis)

T = Tuberous sclerosis

VH3 = Von Willibrand Syndrome

Huntington's chorea

Familial Hypercholesterolemia

Hereditary spherocytosis.



Turner Syndrome

- 60% case - XO
- Webbed neck
- Cystic hygroma
- Lymphedema of hand & feet.
- Primary amenorrhea during puberty.
- Streak ovaries
- Cubitus valgus
- MR rare.
- M/c heart ds - Bicuspid aortic valve & mitral stenosis
↳ half to one-third.
- 20% coarctation.
- Girls infertile

Noonan Syndrome

- AD
- $XX = XY$
- 25% MR
- Valvular pulm. Stenosis.
HOCM
- ASD.

- Girls are mostly fertile.
- Boys - Cryptorchidism.
- Clotting factor deficiency.

Q. Chance of child being not affected if both parents are affected c Achondroplasia is -

A) 0%

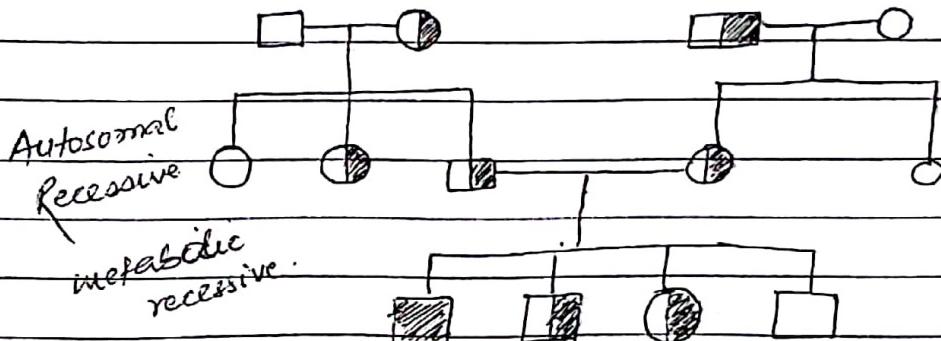
B) 25%

C) 50%

D) 100%

$AAC \times AAC$

(AA) $AAC\ ACA\ ACA^c$





Autosomal Recessive:

Cystic Fibrosis

• δ , AT deficiency

Wilson's ds

Haemochromatosis

Friedreich's ataxia

Gaucher's ds

Niemann's pick ds.

Tay Sach ds.

Hurler's Syndrome.

Mucopolysaccharidosis (MPS)

- Child c noisy breathing
- Coarse facies.
- Ab (n) accumulation of glucoseaminoglycans.
- Gargolism
↳ Chronic rhinitis.

Mucopolysaccharidosis type I → k/a HURLER'S

Syndrome

deficiency of L-iduronidase.

Rx - Enzyme replacement therapy.

HURLER

- MPS-I
- AR
- Corneal clouding

HUNTER Syndrome.

- MPS-II
- XL recessive
- Cornea clear.

Enzyme Replacement therapy:

1st to be treat → ① Gaucher's ds

(β -glucocerebrosidase)



Company name — GENZYME
CER-zyme.

② Pompe's

③ Hurler (MPS-I)

④ MPS-VI (Maroteaux-Lamy)

⑤ X-linked recessive Fabry's

(cns & kidney problem)



Lysosomal disorder.

Cherry red spot macula seen.

Gene therapy:

1st to be treated by Gene therapy → X-linked recessive

Severe combined immunodef. (SCID)

(Adenosine deaminase deficiency)

British guy British lady

Chance of child having cystic fibrosis

UK/Europe → $\frac{1}{25}$ carrier cystic fibrosis.
Ashkenazi Jews

$$\frac{1}{25} \times \frac{1}{25} \times \frac{1}{4} = \frac{1}{2500}$$

$AAC \times AAC$

$(AAC) \cdot AAC AAC AA$
 $\frac{1}{4}$

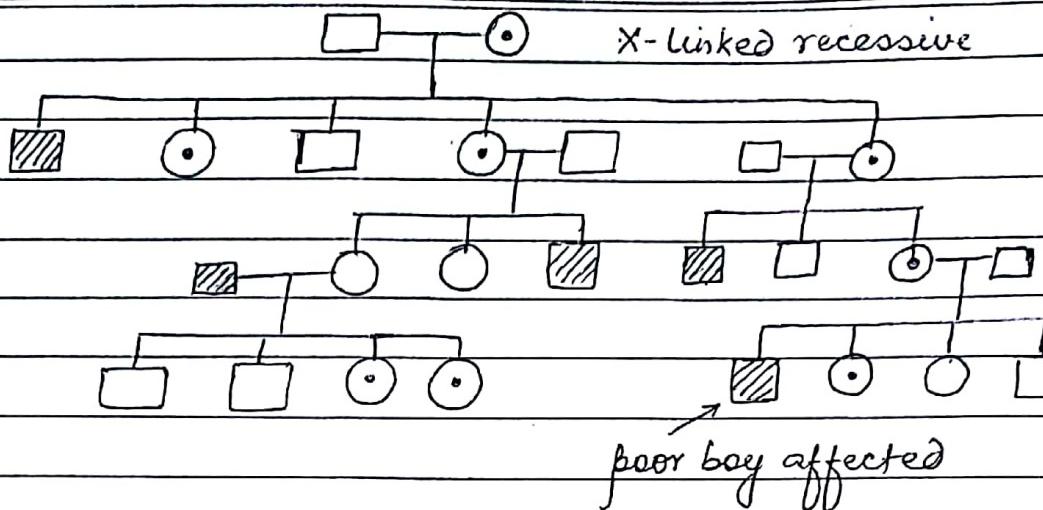
British guy British lady (brother died of cystic fibrosis)

Chance of child to have cystic fibrosis

$$\frac{1}{25} \times \frac{2}{3} \times \frac{1}{4} = \frac{1}{150}$$

$AAC \times AAC$

~~$(AAC) \cdot AAC AAC AA$~~



XX^C XY
 XX XX^C $X^C Y$ XY

X-linked Recessive: (Poor ~~boys~~ boys)

- Duchenne muscular atrophy. (M/c hereditary Neuromuscular disease)
- Hemophilia A & B.
- G-6PD deficiency.
- Wiskott-Aldrich syndrome.
- Colour blindness.
- Lesch-Nyhan syndrome.
- Chronic granulomatous disease.

Duchenne muscular hypertrophy (DMH):

- Pseudo hypertrophy of calf muscle
bcz of fat deposition.
 - Proximal muscle weakness
 - Gower's Sign → Not specific.
- △ → CPK = 10,000 IU

Valley Sign ⇒ Hypertrophy of Supraspinatus

↓ Atrophy of Infraspinatus

Also seen in DMH.

↳ Specific for boys who
don't have calf hypertrophy.

Human Genome:

- 30,000 genes.

Largest gene - *Dystrophin*

↳ Skeletal muscle

↳ Heart - Cardiomyopathy.

↳ Brain → $\frac{1}{3}$ cases MR.

Boys - Duchenne - die teens due to recurrent chest infection.

Duchenne → XLR, $\frac{1}{3}$ de novo mutation.

Becker's dystrophy:

- Similar to Duchenne
- Mild form & present late.
- X-linked recessive.

Wiskott-Aldrich:

- X-linked recessive
- Eczema
- Thrombocytopenia
- Immunodeficiency.

Chronic granulomatous disease -

- X-linked recessive
- Immunodeficiency.
- ~~NADPH~~ NADPH oxidase deficiency
- Dx → NBT dye test

Lesch-Nyhan Syndrome:

- X-linked ~~recessive~~ recessive
- Purine defect
- HGPRTase deficiency.
- Hyperuricemia $> 6.5 \text{ mg/dl}$
- Self-mutilation → nose, palate, fingers.



Q. A mentally challenged child has dysphagia & opisthotonic spasms. He is also having choreoathetoid movements & self mutilation behaviour & no family history. Which of the following investigation is suggested?

- A) Serum uric acid. → Lesch Nyhan Syndrome.
- B) S. ALP
- C) S. LDH
- D) Lead level in blood

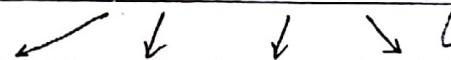
Q. A male child has Fanconi Syndrome & nephrocalcinosis have a variant of dent disease. All true except

- A) Hypercalcioria → 24hr urine $\text{Ca}^{2+} > 4 \text{ mg/kg}$
- B) Proteinuria → LMW (β_2 microglobulinuria)
- C) Similar presentation in father
- D) Rickets

Urolithiasis

Nephrocalcinosis

Fanconi Syndrome - Proximal tubular defect



Na^+ HCO_3^- PO_4^- AA, Glucose

60% >85% >88% 100%

[Lowe Syndrome] - X-linked recessive.

↳ Oculo-cerebro renal gene. (OCRL gene).



Congenital cataract Microcephaly Fanconi's
" glaucoma MR syndrome.

Fructose-1-phosphate] proximal tubular toxic.
Glucose-1-phosphate]

Franconis Syndrome:

- Genetic

↳ X-linked recessive - lowe

XLR Dent

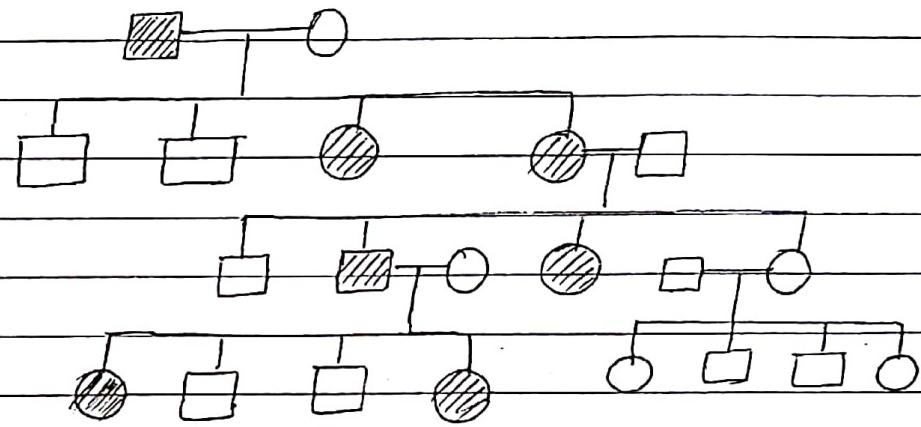
AR

↳ Cystinosis

- Metabolic - Galactosemia, HF-1

Tyrosinemia (R_x - Nifescinac).

- Acquired - expired tetracycline



X-linked dominant father to
all daughters - none son.

X-linked dominant: Males are more severely affected

- Familial hypophosphatemic rickets.

- Urea cycle defect due to OTC deficiency.

- Incontinentia pigmenti [Only seen in girls.]

- Rett's syndrome.

bcz boys die.

(lethal in male fetus).



Rett's Syndrome:

- Pervasive ~~perma~~ developmental disorders.

↳ Autism (Common in boy ~~girl~~ < Syn)

Asperger
Rett's

Q. Not seen in autistic ~~disorder~~ disorder -

- A) Social avoidance
- B) Visual impairment.
- C) Interest in one self
- D) Introvert person.

Asperger's Syndrome: Common in boys

Very good IQ.

Rett's Syndrome - Common in girls

X-linked dominant.

Normal till 6-18 months

Decrease in head growth.

Microcephaly; MR

Hand movements

Repetitive behaviour

MeCP2 gene mutation

↳ Macrocephaly not seen.

↳ Abnormal dendritic morphology in cortical pyramidal cells
(postmortem brain biopsy).

↳ Seizures.

Cause of death → Arrhythmias (cardiac)

↓
Sudden death.

Ornithine Transcarbamoylase deficiency:

Orotic aciduria → URACIL

→ X-linked disorder.

- The mother of these child have also high Uracil level in urine.

Deletions:

Major deletion

→ Cri-du-chat

↳ Deletion of 5p chr.

- Cry like cat due to absent larynx.

Microdeletion

→ Williams (7q23-) Syndrome

→ Prader-Willi Syndrome

(11q11-13-)

→ Di-George's Syndrome (22q11-)

Δ → by FISH.

Williams Syndrome:

Supravalvular aortic stenosis

↳ peripheral pulm. stenosis.

- Hypercalcemia

- Elf in facies.

Di-George's Syndrome:

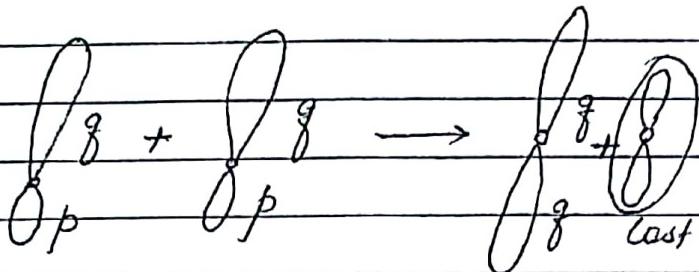
- Hypoplasia of 3rd & 4th parapharyngeal pouch.

- Absent ↙ Thymus

Parathyroid.



TRANSLOCATION:



- Unbalanced

- Robertsonian Translocation :

Translocation b/w two Acrocentric
chromosome.

M/c/c (Genetic) of MR = Trisomy 21 (Down's Syndrome)

Extra chromosome is of females.

Trisomy 21 - In 95% cases : Maternal
meiotic non-dysjunction.

- 3-4% - Robertsonian translocation

- 1-2% Mosaic 47/46

carrier of RT look normal

but they can have abn children.

MOSAICISM:

A single zygote giving rise to different cells.

CHIMERA - different zygote giving rise to different cells
↳ Rare in humans.

- Mosaicism Seen in humans

- 1-2% are down's

Klinefelter Syndrome → 80% XXY; XY/XXY;
XY/XXXYY.

Turner's Syndrome:

Cytogenetics

60% 45X0

15% Mosaic XX/X0

10% Isochromosome Xq or Xp. — Mentally Retarded.

10% 46X deletion.

5% Mosaic X0/XY → Risk of Gonadoblastoma.

Loss of one arm & duplication of other



Isochromosome

Mosaicism ↗ Somatic — Not transmitted

Germline — Transmitted

↳ Blood DNA is normal.

e.g.: ~~Osteogenesis~~ Osteogenetic imperfecta.

- Q. Couple has two children c. tuberous sclerosis. On detailed clinical & lab evaluation (including molecular studies) both parents are normal. ↳ one of the following explains the 2 affected children in this family —

A)

B)

C)

✓ D) Germline mosaicism.

- # Maternal inheritance → Mitochondrial.

Mitochondrial inheritance:

- MERRF (Myoclonic epilepsy & red ragged fibres)
- Mitochondrial encephalopathy, stroke-like episodes, & lactic acidosis (MELAS)
- Leber hereditary optic neuropathy (LHON)



- Leigh disease
- Kearne - Sayre Syndrome (KSS) (ophthalmoplegia)
- NARP (Neuropathy, Ataxia, Retinitis pigmentosa)
- Chronic progressive ophthalmoplegia
- Pearson's Syndrome : Panhypoproteinemia
+ Pancreatic insufficiency

Anticipation : Severity of genetic disorder ↑ in every successive generation.

More repeats — more problem.

eg: All Tri nucleotide repeat disorders

- Fragile X — CGG repeats
- F. Ataxia — GAA "
- Myotonic dystrophy — CTG, CCTG
- Spinobulbar muscular dystrophy — CAG
- Huntington's — CAG
- Spinocerebellar ataxia — CAG/CTG.

Fragile X : genetic

- End w/c cause of MR in boys.
- X-linked
- Large face
- Large ears
- Prominent jaw
- Large testes..
- CGG repeats > 1500 repeats

Genomic imprinting:

Chr. 15q • 11-13 deletion.

↳ Paternal inheritance cause Prader-Willi Syndrome
↳ Maternal " " " Angelman "

- Differential expression of genetic appearance depending upon parents.

Prader-Willi Syndrome

- Severe neonatal hypotonia.
- Obesity
- Small hands & feet. (Rx - GH treatment)
- Unusual behaviour
- MR.
- High Ghrelin → So obese. (Rx - Anti Ghrelin)

Angelman Syndrome:

- (R) at birth
- K/A happy puppets.
- Subsequently develop seizures
- MR
- Ataxia

↓
appetite hormone
for foodies.

Genomic imprinting → Prader Willi Syndrome (70%)

↓

Almond shaped eyes.

② Angelman Syndrome (70%)

③ Neonatal DM

④ Beckwith Wiedemann

⑤ Seckle

⑥ Temple

⑦ Wang

⑧ Pseudohypoparathyroid Ib.



Q. Father carrier of cystic fibrosis - AAC^c
Mother - $(N) \rightarrow \text{AA}$



Yes there is a chance of cystic fibrosis in child
 ACAC^c



Uniparental disomy.

- In some cystic fibrosis
- In some sickle cells.
- 30% Prader Willi - Unimaternal disomy.
- 5% Single man - Uniparental disomy.

PRADER WILLI :

70% - Paternal inheritance deletion.

30% - Unimaternal disomy

↳ Maternal silencing gene.

Congenital Heart de:

NADA'S CRITERIA:

1 Major & two minor criteria at least.

Major:

- ① Systolic murmur grade III or more ass. c thrill.
- ② Diastolic murmur
- ③ Cyanosis (Central)
- ④ CHF

Minor:

- ① Systolic murmur < grade III
- ② Abn S₂
- ③ Abn ECG
- ④ Abn X-Ray
- ⑤ Abn BP

A₂ ----- P₂ Expire

A₂ ----- P₂ Inspire

ASD - Wide & fixed S₂.

A₂ ----- P₂ Expire

A₂ ----- P₂ Inspire.

- Volume overload in right ventricle.

VSD - Wide & Variable S₂.

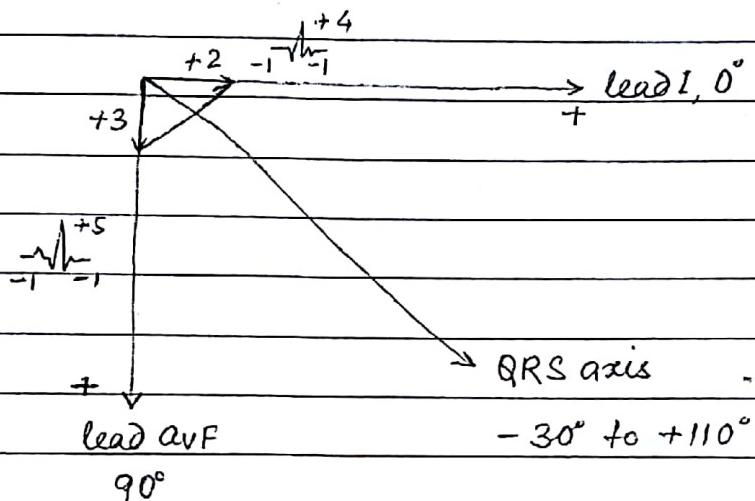
A₂ ----- P₂

A₂ ----- P₂

TOF - Single S₂ (A₂).

All newborn has RVH & RAD.

- Axis like adult > 1st month life.
- T-wave V₁; V_{3R}; V_{4R}
- 'up' first 48 hrs.



- Negative after 48 hrs
- Never be +ve < 6 yrs
- > 6 yrs → positive.

Prevalance of CHD:

- Prevalence = 0.8 - 1%

Recurrance = 2-6%

M/C CHD = VSD (30-35%).

2nd M/C CHD = ASD (Secundum) - 6-8%
 > PDA (6-8%)

> Co-arctation of Aorta (5-7%)

> TOF (5-7%)

> Pulm. Valve stenosis (5-7%)

> Aortic * * (4-7%)

M/C Syndrome in CHD = Down's Syndrome.

Down Syndrome:

M/C → Complete AV septal defect (CAVSD) /
 AV canal defect / Endocardial cushion defect
 Ostium primum ASD.

(37%) — M/C/c of death.

- VSD (31%)
- ASD (15%) → Secundum
- Partial AV septal defect (PAVSD) — 6%
- TOF → 5%
- PDA → 4%
- Miscellaneous → 2%

Turner's → half to one third bicuspid aortic valve;
 20% coarctation!

Noonan's → Turner's phenotype.

AD; XX = XY

Valvular pulm. stenosis.

HOCM; ASD.

Rubella → PDA; Peripheral pulm. stenosis; VSD.

↓
 Rubella

Alagille Syndrome
 William's "

Maternal lithium → Ebstein's anomaly.

Maternal mumps → Endocardial fibroelastosis/
 LV obstruction newborn.

Maternal penicillamine → Cutis Laxa.



Maternal SLE << Maternal Sjogren Syndrome

- New born complete heart block.

pacemaker insertion (anti ~~Ro~~ Ro).

Maternal warfarin:

Chondrodysplasia punctata.

Maternal thalidomide: Phocomelia.

Foetal circulation:

- 1 Umbilical Vein (left)
- 2 umbilical artery.

Ductus venosus →

P_{O_2} in umbilical vein = 30-35 mm of Hg

P_{O_2} of IVC in fetus = 28-30 mm of Hg.

As soon as child takes his 1st breath

- Umbilical artery constricts.



U. vein closes



Ductus venosus closes.

Ductus arteriosus:

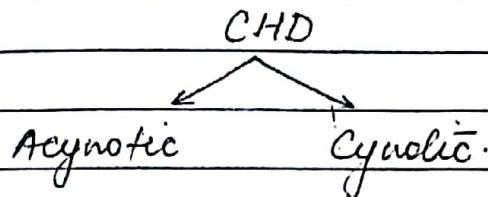
- Physiological closes in 10-15 hrs.

- Anatomical → 10-21 days.

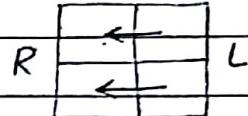
Foramen ovale:

Functionally closes by 3 months.

Anatomically 10-15% open.



Acyanotic : $L \rightarrow R$ Shunt
ASD, VSD, PDA.



Pulm. blood flow ↑ (Plethora)

↳ lung vascularity is good.

C/F : Failure to thrive

Recurrent pneumonia

Feeding diaphoresis

CHF in 6-10 wks of life.

Suck-rest-suck cycle

↑ sweating (d/t sympathetic stimulation).

Tachycardia] in every CHD.

Cardio megaly])

Cyanotic

PBF

↑

↓

Plethora

Oligemia

- Persistant Truncus

arteriosus

(Cyanosis + Recurrent pneumonia)

- dTGA + VSD

- TAPVC

Massive

Cardiomegaly.

· Ebstein's

(Box/Ballot heart)

physiology.

① Heart
Single S₂

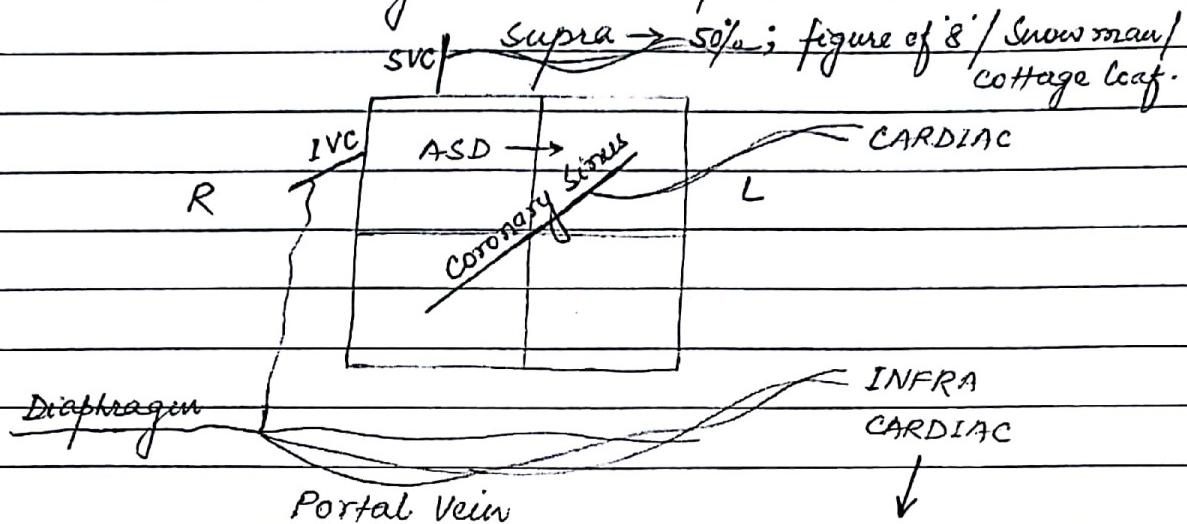
F TOF

L DORV + PS

O dTGA + VSD + PS

T Single Ventricle + PS.

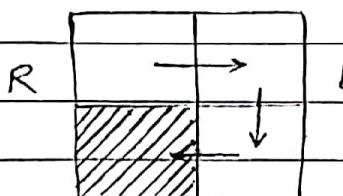
TAPVC (Totally anomalous pulm. venous connection):



More severe TAPVC -

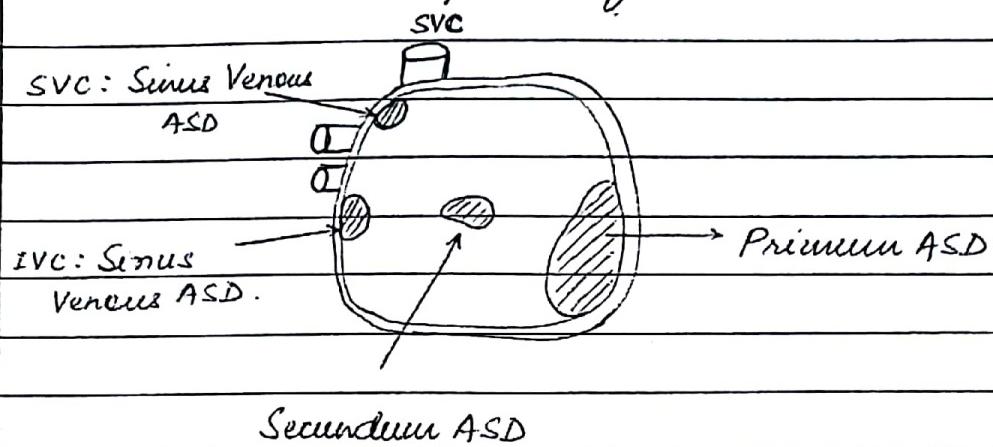
- Cyanosis at birth.
- Obstructive pulm. venous hypertension.
- Ground glass : Kerley B lines.
- = - Worsen by PGE1 infusion.
- Only pediatric cardiac Sx emergency.

Tricuspid Atresia:



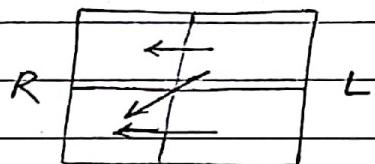
- Cyanosis + ↓ PBF
- + LVH
- LAD

ASD (Atrial Septal defect) :



M/C syndrome ass. = Down's Syndrome.

Endocardial cushion defect.



- Very large L-R shunt.
- PBF↑
- Pulm. plethora

ASD Syndrome :

- HOLT ORAM : • Familial, AD,
 - ASD Secundum; VSD; 1 degree block;
 - AF + Bone defect (Absent Radius).
 - Distally placed thumb / Rudimentary thumb / Triphalangeal thumb.
 - A/K/A Hand-heart Syndrome.
 - TBX5 mutation → Pleiotropy.
 - ↳ Common transcriptional factor for hand & heart.



Absent Radius associations:

- ECG (Holt - oram)
- Platelet (Thrombocytopenia ; AR).
- Bone marrow biopsy (Fanconi's anemia)



Congenital aplastic anemia

- Rarely Karyotyping (Edward Syndrome)

ASD Syndromes:

- Down's Syndrome
- Holt oram
- Lutembacher → ASD + Mitral Stenosis
- Ellis Van Creveld → ASD + polydactyly.

ASD Secundum:

- Child → asymptomatic, wide & fixed S₂.
- ECG → RAD (Right axis deviation).
- In adult life → Complications
 - RV failure
 - Arrhythmias; AF → CVA
 - Reversal; R → L (Eisenmenger Syndrome)

Natural history:

If < 3 mm → close itself

> 8 mm → unlikely to close; Require Sx.

Indication of Sx in ASD Secundum:

- All symptomatic.
- Q_p / Q_s > 2 ; even if no symptoms.
 ↳ Pulm blood flow / Systemic blood flow.

Q. Least chances of Infective endocarditis is seen in ?

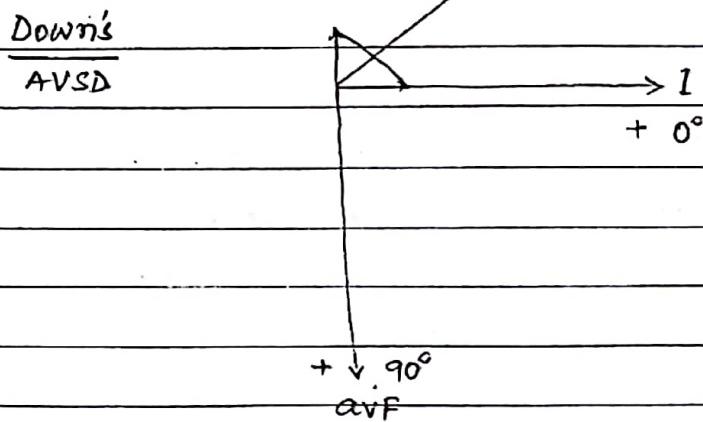
- A) Small VSD \rightarrow M/c/o of IE
- ~~B) Small ASD secundum \rightarrow Rare~~
- C) Mild AS
- D) Mild AR.

ASD Secundum doesn't require any prophylaxis t/t before going to Sx.

ASD primum + Mitral Regurgitation:

- Wide & fixed S₂ +
 $S_1 = S_2$ apex \rightarrow axilla & back.
- 6 - 10 wks presents in CHF.
- Conduction defects ; ECG \rightarrow LAD.
- Common in Down's Syndrome baby.

LAD d/t endocardial cushion defect.





VSD (Ventricular Septal defect):
70% → Perimembranous

Small VSD

- Root of aorta $< \frac{1}{3}$
- $< 3 \text{ mm}$
- Called Maladie de Roger's defect.
- Loud murmur (S₁) (Pansystolic)

↓
lower left Sternal border.

- Asymptomatic

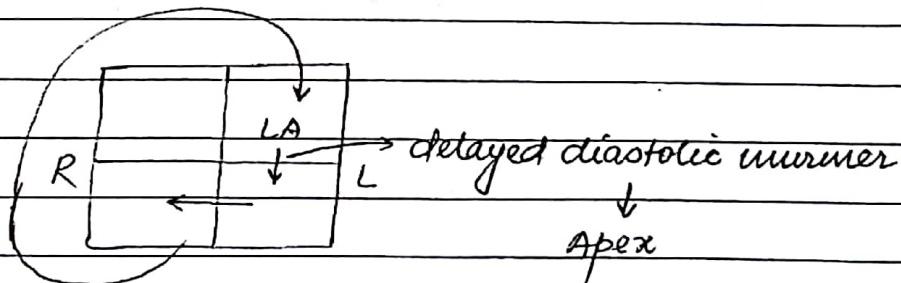
Medium VSD



CHF at 6-10 wks of life

- No murmur/
- Ejection systolic murmur.
(b/w S₁ & S₂).

In medium to large VSD, Left Atrium enlarges first.
In Small to medium VSD, left Ventricular hypertrophy.
due to blood overload.



Natural course of VSD closure:

- 80% perimembranous
- 50% muscular VSD's close.
- By 4 yrs.

Indication of Sx in VSD:

- Failure of Medical therapy.
 - ↳ Digoxin
 - ↳ Diuretics
 - ↳ Dilators
 - even ↳ ACE i (Remodelling of heart).
- $Q_p/Q_s > 2$; if no symptoms.
- Swiss cheese VSD. (Multiple; apex)
- Supracrestal (outflow)

Large L → R shunt



Pulm. blood flow ↑



Irreversible changes pulm. microvascular.

[Pulm. HTN]



RV pressure ↑; RVH



Reversal; R → L (Eisenmenger Syndrome).



Sx is C/I !

Differential clubbing: Toes > fingers



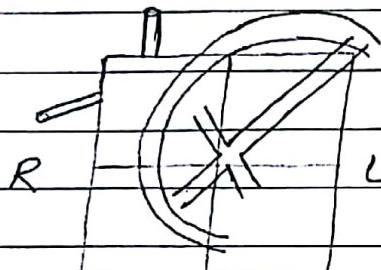
PDA + Reversal.

(also differential cyanosis).

Down's Syndrome baby c Endocardial cushion defect undergoes Eisenmenger Syndrome.



Ductal dependent lesion:

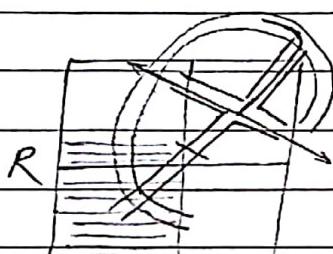


- S/E - Apnoea.
- HLHS \Rightarrow Rx - (PGE₁) infusion.
- Critical AS
- Preductal coarctation
- Interrupted aorta
↳ Shock.

I > Systemic blood flow dependence.

II > Pulm. blood flow dependence.

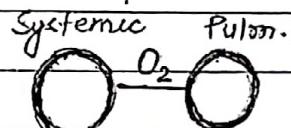
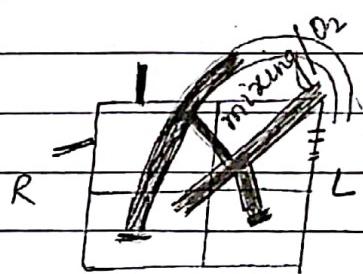
- Tricuspid Atresia
- Pulm. Atresia



↳ Central cyanosis.

In emergency cond'n \rightarrow PGE₁ infusion.

III > d-TGA



Rx - PGE₁ infusion

d-TGA \rightarrow dependent for mixing.

Truncus Arteriosus is Ductal independent.

Emergency Sx in TGA \rightarrow Rashkind's
Balloon atrial septostomy.

Definitive Sx \rightarrow Jatene's Arterial Switch.
↳ Best time: c in first
2 weeks of life.

M/c Cyanotic Heart de:
 Overall - TGA

In Infants - d-TGA (< 1 yrs)
 > 1 yr - TOF

Hyperoxia test in cyanotic newborns -

- 10L O₂ to cyanotic newborn
- $pO_2 > 150$ mm Hg \rightarrow excludes heart de.

① 50% d-TGA + VSD \rightarrow Mild

② d-TGA + intact septa (complete d-TGA) - Birth.

③ d-TGA + patent foramen ovale \Rightarrow At birth.

$$\textcircled{1} < \textcircled{3} < \textcircled{2}$$

TOF (Tetralogy of Fallot):

M/c cyanotic heart de beyond infancy.

- ① Narrowing of the pulm. valve (infundibular).
- ② RV hypertrophy.
- ③ Overriding of aorta over VSD.
- ④ VSD - opening b/w left & right ventricles.
 (perimembranous).

Pink child become a blue TOF.

- Cyanosis; clubbing.
- Polycythemia \rightarrow R/F of Renal vein thrombosis (RVT)
- Hematuria, ~~microhematuria~~. Flank mass, Anemia.
- Infant - Cyanotic/tet/hyperpneic spells \rightarrow older squat
- Complications:
 $\textcircled{1} < 2$ yrs = thrombosis



(ii) $> 2 \text{ yrs} = \text{Brain Abscess}$.



in the territory of middle
meningeal artery
(Parieto - Temporal)

Infants \rightarrow Cyanotic / tet / hypoxic spells — older squat

(R : knee chest position; — Systemic Vascular Resistance falls.

i.v. Ketamine; — R \rightarrow Shunt ↑ — Murmur disappears.

Phenylephrine) — PO₂ falls (R : O₂; i.v. NaHCO₃).

- Respiratory centre + + + (R : S/c Morphine)
- Dynamic Pulm. stenosis (R : i.v. Propranolol)
- Crying (Bcoz of ↑ venous return).

In TOF — loud Holosystolic murmur at the left
2nd - 3rd ICS due to pulm. stenosis.

Palliative Shunt

- Modified Blalock Taussig \rightarrow Subclavian
to Pulm. artery
(opposite to Aortic Arch.)
- Waterston
- Pott's (Descending Aorta to left pulm. artery)



25% TOF \Rightarrow has Right aortic arch.



M/c c 50% of Truncus arteriosus

Pentalogy = + ASD

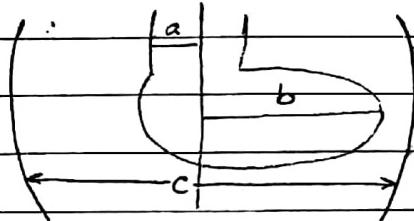
Trilogy = Overriding of aorta absent; VSD absent; ASD present.

[CHF never seen in TOF

Cardiomegaly never seen in TOF.

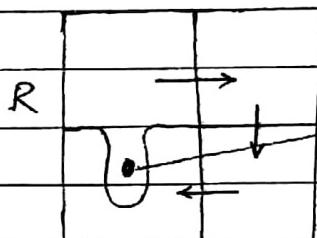
↓
Cardiothoracic ratio > 0.6 infant

> 0.55 in older infant.



"Boot shaped heart".
"Coer en Sabat".

Ebstein's anomaly:



• Cyanosis \Rightarrow ↓ PBF

• Maternal lithium

→ Atrialisation of RV.

Pressure = RA

Ventricular = ECG

disconcordance b/w pressure &
ECG.

• Intracardiac ECG helpful in Δ.

• Systolic; diastolic murmurs.
Quadruple rhythm

• Box/Ballon; Massive heart.

• SVT; WPW Syndrome.



Q A neonate presents with recurrent abdominal pain, restlessness, irritability & diaphoresis on feeding. Cardiac auscultation reveals a non-specific murmur. He is believed to be at risk of MI.

The most likely Δ is —

- A) VSD
- B) ASD
- C) TOF

D) Anomalous origin of the left coronary artery
= ACCAPA

- Q wave in Lead I; aVL
- LAD branch absent.
- Ischemic LV → Anterolateral MI.

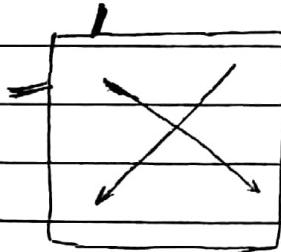
On Angiography → Rt coronary artery fills.
- Lateral MI in infant.

Rx: Bypass Sx.

l-TGA → Corrected TGA.

Normal Heart.

Atrio-Ventricular discordance.



[RA connected to LV
LA " " RV]

No problem to baby.

KAWASAKI'S DS:

- M/c acquired heart disease in US, Japan, & Chandigarh.
- Fever $>$ 5 days.
- Development of a limp.
- Erythematous macular exanthem over body.
- Ocular conjunctivitis.
- Dry & cracked lips.
- Red throat & cervical lymphadenopathy.
- Grade II/IV vibratory systolic ejection murmur at lower left sternal border.
- Predominant Neutrophils & ↑ platelet.
- M/c medium sized vasculitis.
- 20-25% causes develop aneurysm in future.

Q. Which of the following vasculitis not occur in adults?

A) Kawasaki's (85% are < 5 yrs)

B) Sjögren's Syndrome - Seen in adult females.

C) Giant cell arteritis

D) HSP

Peeling of palm & soles is classically seen in 2nd - 3rd wk of life.

Rare in B. Before 3 months of life.

DOC : IV Ig.

Δ: Fever $>$ 5 days & any 4 of these 5

- ① Changes in extremities (eg: Erythema, edema, desquamation).
- ② B/L conjunctivitis (not ass. w/ exudates)
- ③ Polymorphous rash (not vesicular)
- ④ Cervical lymphadenopathy.



⑤ Changes in lips & Oral cavity (eg: Pharyngeal erythema; dry/fissured or swollen lips, strawberry tongue).

Non-classical feature of Kawasaki:

Arthralgia; Arthritis;

Thrombocytosis; Urethritis;

aseptic meningitis (irritable)

M/C/C of death in Kawasaki —

Overall — Coronary artery aneurysm

Acute phase — Myocarditis.

Rx: 2v Ig 2g/kg in acute phase reduces risk 4-6%

Aspirin 100mg/kg/day X 2 weeks.

If Resistant to Ig: 10-15% cases

↳ Add steroid (Methylprednisolone).

↳ Repeat Ig

↳ TNF blockers → Infliximab; Etanercept.

↳ IL-1 inhibitor → ANAKINRA

Recurrence Rate → 1-2% cases.

Mx of aneurysm:

Small → 50% resolve over 1-2 yrs.

Aspirin 3-5mg/kg/day.

Medium to large → Add Warfarin

Sx (may be bypass)

HSP (Henoch - Schönlein Purpura) :

M/c vasculitis (small vessel) Overall .

M/c leucocytoclastic vasculitis .

- Palpable non-thrombocytopenic purpura .
- IgA deposition of vessels in dermis .

Pathogenesis : Aberrant Galactosylation

Mesangio-proliferative disorder -

Small vessels → Skin

Arthritis

Arthralgia

GIT - mesenteric ischaemia

Kidneys - HSP Nephritis (40-50%).



84% develop in 4 wks

91% " " 6 wks

97% " " 6 months .

- Microscopic haematuria

- Proteinuria

- 1-2% RPGN (over days to week)

↳ On Biopsy crescent seen

↳ ∴ Crescентic GN



proliferation of parietal
epithelial cells .

Rx : i.v. Methylprednisolone .



Rheumatic Fever:

- M/c acquired heart ds in India / Developing country.
- Due to Group A β -hemolytic streptococci strains M-type 1, 3, 5, 6 & 18.
- Most frequent b/w 5-15 yrs.
- Latent period 3 weeks.
- Autoimmune : Molecular mimicry

Affects ↗ Myocardium
Muscle.

Modified Jones (2015):

- Era of Echocardiography \rightarrow Subclinical AR/MR.
- A/c to Risk area.

Low Risk - defined as having an ARF incidence < 2 per 100000 school-aged children.
(usually 5-14 yrs old) per year or an all age prevalence of RHD of ≤ 1 per 1000 population per year (Class IIa ; level of evidence C).

Criteria = 2 major or 1 major + 2 minor
+ Essential criteria.

Recurrence = 2 major or 1 major + 2 minor or
3 minor

Major Criteria:

Low risk populations:

- Carditis (clinical &/or subclinical)
- Arthritis (Polyarthritis only).
- Chorea
- Erythema marginatum
- Subcutaneous nodules.

Moderate to high-risk populations:

- Carditis (clinical &/or Subclinical)
- Arthritis
 - Monoarthritis or polyarthritis
 - Polyarthralgia \oplus
- Chorea
- Erythema marginatum
- Subcutaneous nodules.

Minor Criteria:

Low risk populations:

- Polyarthralgia
- Fever ($\geq 38.5^{\circ}\text{C}$)
- $\text{ESR} \geq 60 \text{ mm in the first hour}$ &/or $\text{CRP} \geq 3.0 \text{ mg/dL}$
- Prolonged PR interval, after accounting for age variability.

Moderate to High risk populations:

- Monoarthralgia
- Fever $\geq 38.5^{\circ}\text{C}$
- $\text{ESR} \geq 30 \text{ mm in first hour}$ or $\text{CRP} > 3 \text{ mg/dL}$.
- Prolonged PR interval - Also in MR & MS.



Rx:

- Aspirin 100mg/kg/day x 12 wks.
- Prednisolone x 12 wks.
 - 2mg/kg x 3wks → taper
 - Severe carditis or CHF.
- Crystalline Penicilline G x 10 days
- IE prophylaxis.

Mitral valve → M/c involved in RF

↳ Recurrence cause Mitral stenosis.

↓
Primary prevention

In India 2^o prevention

↳ Penicillin G

Benzathine (i.m.)

- 600,000 IU for children wt ≤ 60lb

1.2 million IU for children wt > 60lb
every 4 wks.

or, Penicillin V (Oral)

↳ 250 mg twice a day

or, Sulfadiazine or Sulfisoxazole (oral)

↳ 0.5g once a day for pt. wt ≤ 60lb

1.0g once a day for pt. wt > 60 lb.

For pt. allergic to penicillin & Sulfa group.

↓
Macrolides are given orally.

CATEGORY	DURATION
- Rheumatic Fever \ominus out Carditis	5yr or until 21 yrs of age whichever is longer.
- RF \ominus carditis but \ominus residual heart ds (No valvular ds)	10yr or until 21 yrs of age, whichever is longer.
- RF \ominus carditis & Residual heart ds (persistent Valvular ds)	10 yrs or until 40 yrs of age whichever is longer. <i>Sometimes lifelong prophylaxis.</i>

Q

- Blood pressure = 86/600 mm Hg

- 4 yr, unconsciousness.

- HR = 180/min

- CFT = 4 sec

$\Delta = ? \rightarrow$ Compensated Shock.

Rx = 20 ml/kg 0.9% NaCl.

SHOCK = BP < 10th centile for age & sex.

Criteria for Shock:

Criteria for Hypotension by age.

Age	Systolic BP
Term neonates (0 to 28 days)	< 60 mm Hg
Infants (1-12 month)	< 70 mm Hg
Children (1-10 yrs)	< 70 + (age in yrs X 2)
Children > 10 yrs	< 90 mm Hg

Compensated shock: Rx - 20 ml/kg 0.9% NaCl

Repeat upto 60 ml/kg

↳ CVP line (next step).

• low - fluid given.



- Normal → Cold epinephrin
- =
- Warm: Nor-epinephrin.

Hypertension $BP > 95^{\text{th}} \text{ centile}$

- Essential ~10% ; increasing
- Secondary HTN

↳ Renal parenchymal - Reflux Nephropathy
VUR

↳ Renovascular

• Major → RAS; RVT

Minor → HUS

↳ Cardiac - post ductal coarctation (Tunne)

↳ Endocrinol - • Hyperthyroid

• Cushing

• Pheochromocytoma.

• CAH 11 beta/17 alpha

hydroxylase deficiency

End organ damage in HTN :

- Fundus
- Echocardiography concentric LVH.
- Any adolescent $BP > 130/80$
In children $> 10.4 \text{ yrs} \rightarrow 120/80 \rightarrow \text{HTN}$
- Urine protein.

Rx : HTN

- Life style modification.
- Pharmacological therapy - end organ; symptoms;
↓ Severe $> 99^{\text{th}} \text{ centile}$.

ACEi / ARBs.

ACEi C/I if GFR < 30 → causes hyperkalemia

↓
Rx → Amlodipine (ccb's)



Date _____

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Page _____

Hypertensive Emergency:

- LVF \Rightarrow S₃; Gallop; Basal crepts.
- Seizures.

R:

Best: i.v. Nicardipine infusion

R: Sodium Nitroprusside i.v./

Esmolol /

Labetalol i.v.

Linezolid, Nitroprusside & Amphotericin B
should be covered.

Q. A 12yr old boy c seizure.

BP = 200/140.

Femoral pulses not palpable.

$\Delta = ?$

A) Takayasu aortoarteritis (R- Prednisolone).

B) Grand Mal Seizures.

C) Fibromuscular dysplasia (FMD)

D) Renal parenchymal defect.



Tachyarrhythmias

- Based on QRS.

~~wide QRS~~

Wide > 0.09 sec (VT/VF)

- Pulseless

- Stable

- Arrest.

Rx - lignocaine

Rx - Defibrillation $0.5 - 1 J/kg$

Amiodarone

Narrow

- Recurrent SVT

- HR > 220 infant; > 180 older

- P-wave absent; inverted

Vagal maneuvers

Stable

Rx - Adenosine (fastly given \pm saline flush)

↓

as close to the heart
as possible.

CHF; Shock

Rx - Synchronized cardioversion

Respiratory

Cough & cold caused by Rhinovirus.

JMNCS (7/L)

Age = 2m - 12 months = RR \approx 50 or more.

Age 12m - 60m = RR \approx 40 or more.

- Chest is Indrawing = Pneumonia.
- or fast breathing

Rx :- Give oral Amoxicillin for 5 day.

Any general danger sign \Rightarrow Severe Pneumonia.
or
Stidor in calm child } or very severe dz

R

- Give 1st dose of an appropriate A/b.
- Refer to hospital.

Q. 18 m old child weighing 11.5 kg comes to PHC with resp. difficulty. O/E lethargic RR = 46/min. No chest retraction = Next Step.

Ans (b) Prescribe oral antibiotic & Refer to higher centre.

→ Signs (< 2 months)

Next step

- Convulsions or

Serious = Give i.v.

- Fast breathing (≥ 60)

Bacterial \Rightarrow Gentamycin.

- Severe Chest Indrawing.

Sug®

- 10 or more skin pustules.

↓
Refer to
higher centre

or a big boil or

If axillary temp 37.5°C

or above or less than 35.5°C

- Drowsiness / Unconsciousness

- less than R.R limit.

- Umbilicus red or draining \rightarrow local (= Give oral)
- Plus) pus discharge from ear or skin pustules. $\left.\begin{array}{l} \text{ear} \\ \text{skin} \end{array}\right\}$ Back $\left.\begin{array}{l} \text{ear} \\ \text{skin} \end{array}\right\}$ Suf^e

- Causes of Pneumonia Age wise in India & world
- Neonates = Grp. B streptococcus, E. coli.
< 3 weeks.
- 30K - 3m = Respiratory Syncytial Virus (RSV)
S. pneumoniae, H. influenzae.
- 4m - 4yrs = RSV, S. pneumoniae, H. influenzae.
 $> 5 \text{ yrs}$ = M. pneumoniae, S. pneumoniae.
- H. influenzae $\xrightarrow{\text{dt}}$ Vaccination $\rightarrow \downarrow$ in Incidence.

Viral pneumonia

- Prodrome.
- Diffuse, Bl
- Not lobar pneumonia
- Interstitial infiltrate

T/E: ① Doc for RSV = Ribavirin.

② Influenza = Amantadine, Rimantadine

H1N1 = Influenza A \rightarrow Neuraminidase Inhibitors.

\hookrightarrow Oral Oseltamivir.

\hookrightarrow Inhalated Zanamivir.

Pandemic :-

Phase 1 - 3 Animals.

Phase 4 - May 27, 2009 (Mexico)

\hookrightarrow Human-human transmission at community

Phase 5 - Aug 5/09 DSA.

Human - Human transmission in one zone

Phase 6 = June 11, 2009, US & India.

→ Human - human transmission in two zones

H₁N₁ Virus :-

- SS RNA.

- Belongs to Orthomyxoviridae.

- Size = 80 - 200 nm.

Types :- A, B, C.

Surface Ag = H (Hemagglutinin)
N (Neuraminidase)

As the reassortment occurs in swine flu.

= Swine flu.

D/T Antigenic shift & drift - No vaccines made

Symptoms = Flu like.

(Complications of H₁N₁) → in High risk groups.

⇒ Pneumonia (viral)

⇒ Bacterial Superinfection.

⇒ ARDS like features.

High Risk groups :-

Comorbidity.

→ They were associated with some mortality

→ Pregnancy

→ Nephrotic syndrome, Chronic illness, post transplant

→ <1 yr., >65 yr.

Indications of oseltamivir

① Give it to all suspected cases of H₁N₁.

② Give it to all confirmed case.

ASis by throat swab & Nasopharyngeal.

Swab I send for Real time PCR.

③ H₁N₁ influenza.

④ Give it to all household contacts, occupational

adults

(Post exposure in doctors) -

Dose = 75 mg (op BD - 5d).

Prophylactic dose = 75 mg OP → 7-10 days -

zamavir.

The dose = Two 5mg Inhalations (10mg total)
twice / day x 5 days.

Prophylaxis dose = twox5mg Inhal^(R) - 0).

Q:- H₁N₁ 10 wks pregnant -

→ Give oseltamivir [NOT teratogenic]

→ T₂

[↳ only in rats]

Q:- Pt on Rifampicin Induces hepatic CYP450.

gets H₁N₁ sy^(R)

→ What happen to dose of oseltamivir

dose = Remains same → Renal ex^(R)

Q:- Dose in hemodialysis

= oseltamivir & dose is reduced

Pneumonia

- St. pneumonia = causes severe, lobar pneumonia.

Incub/ Rd = 1-3 days

(Rx) } Ceftriaxone, DOC.

→ Resistant Variety = DOC ⇒ Vanco + ceftriaxone

Vaccine :- > 2 yrs. children = PPV ~ 23.

< 2 yrs = PCV - 7/11 = conjugated.

6, 10, 14 weeks, 1/3 booster at 15-18 months.

IGAVI :- Global Alliance for Vaccines & Immun^(R)

- founded by Bill Gates & Melinda Gates

- free vaccines in poor country

Staph. Aureus

- Max Mortality (10-30 %)
- Ch. - Air filled cavity (Pneumatocele)
 - also seen in d/t.

1) Klebsiella.

2) Kerosene oil poisoning

→ Pneumatocele can Rupture & develop pneumothorax

- S. aureus = 1/3rd of sepsis in children.
(Plus in pleural cavity)

T/t = ICDT.

DOC for S. aureus = Clavacillin.

Vancomycin for MRSA.

- H. influenzae = Usually part of S. sepsis.
- can have arthritis, meningitis.

Rx = ampicillin & chloramphenicol.
20-40% are resistant.

Rx: (Ceftriaxone) → DOC.

Atypical pneumonia

- Rate - < 4 yrs = ~~> 4 yrs~~, > 5 yrs. !
- Symptoms - dry cough.
- Interstitial pneumonia
- Organism = Mycoplasma chlamyde.

Rx: - Macrolides.

15 month

Motor → Walks alone, crawls up stairs

Adaptive → Makes tower of 3 cubes, makes a line with crayon, inserts raisins in bottle

Language -唐语; follows simple commands, may name a familiar object (eg. ball) responds to his/her name.

Social → Indicates some desires or needs by pointing, hugs parents.

18 month

Motor - Runs stiffly, sits on small chair, walks up stair with 1 hand held, explores drawers & waste baskets.

Adaptive → Makes tower of 4 cubes, imitates scribbing, imitates vertical strokes, dumps raisin from bottle.

Language - 10 words (average), names pictures, identifies 1 or more parts of body

Social → Feeds self, seeks help when in trouble, may complain when wet or soiled, kisses parent with pucker.

24 month

Motor → Runs well, walks up & down stairs, 1 step at a time, opens door, climbs on

furniture, jumps.

Adaptive → Makes tower of 7 cubes (6 at 21 months)
scribbles in circular pattern, imitates

H_z stroke, folds paper once imitatively

Language → Puts 3 words together (subject, verb, object)

Social → Handles spoon well, often talk about
immediate experiences, helps to undress,
listens to stories when shown pictures

30 months

Motor → Goes up stairs alternating feet

Adaptive → Makes tower of 9 cubes, makes V & horizontal strokes, but generally will
not join them to make cross, imitates
circular stroke, forming closed figure

Language → Refers to self by pronoun 'I'
Knows full name

Social → Helps put things away, pretends in
play

36 months

Motor → Rides tricycle, stands momentarily
on 1 foot

Adaptive → Makes tower of 10 cubes, imitates
construction of bridge of 3 cubes

~~copies circle, imitates cross~~

Language - Knows age & sex, counts 3 objects
incorrectly, repeats 3 numbers or a sentence of 6 syllables, most of speech intelligible to strangers.

Social → Plays simple games (in parallel with other children) helps in dressing (unbuttons clothing & puts on shoes) washes hands.

48 months

Motor → Hops on 1 foot, throws ball overhead, uses scissors to cut out pictures, climbs well

Adaptive → Copies bridges from ~~parallel~~ model, imitates construction of gate of 5 cubes, copies cross & square, draws man with 2-4 parts besides head, identifies longer of 2 lines.

Language - Counts 4 pennies accurately, tells story

Social → Plays with several children, with beginning of social interaction & role-playing, goes to toilet alone

60 monthly.

Motor - Skips

Adaptive → Draws Δ from copy, names heavier of 2 weights

Language - Names 4 colours, repeats sentence of 10 syllables counts 10 pennies correctly

Social → Dresses & undresses, asks questions about meaning of words, engages in domestic role playing.



Fever & Stridor. Inspiratory sound in upper airway.

Croup / LTB

- 75% parainfluenza

→ Prodrome → Stridor

Barking Cough.

Subglottic narrowing

K/A = Stippled sign

On X-ray:

↳ Doc = formild = dexta = 0.6 mg/kg. mod & severe

Epiglottitis

In world - S. pyogenes.

S. pneumoniae

S. aureus

In India - H. influenza.

dat X-Ray → Thumb Sign.

Rx ^{Neck} → Airway [Emergency tracheostomy]

Ceftriazone + Sulbactam
(ESBL)

↳ Nebulisation & Racemic epinephrine.

→ Westley Croup Score Criteria to dx it.

BRONCHIOLITIS:

- Inflammatory obstruction of smaller airway.

M/c organism - RSV (50%)

↳ Respiratory Syncytial Virus.

R/F:

- Males, Top fed.

① - Preterm ; chronic lung disease.

② - L → R shunt.

- Smoking mothers.

Airway resistance, $R = \frac{1}{r^2} \rightarrow$ air trapping

Prodrome → Wheeze / ~~or~~ Ronchi.

X-Ray chest shows Hyper inflation.

Rx: Antibiotics has no role.

Humidified O₂.

For 2 High Risk group - Nebulised Ribavirin
Palivizumab

Long term - Persistent wheeze in infancy

↓
Reactive airway disease.

ASTHMA

Classification a/c to severity.

	Step	Symptoms	Night symptoms	Peak expiratory flow rate
I.	Intermittent.	<1 time a week; asymptomatic & (N) PEFR b/w attacks	≤ 2 times a month	≥ 80% predicted; Variability <20%
II.	Mild persistent.	>1 time a week but <1 time a day	>2 times a month	≥ 80% predicted; Variability 20-30%
III.	Moderate persistent.	Daily use β -agonists; daily attacks affect activity.	>1 times a month	>60% & <80% predicted; Variability >30%
IV	Severe persistent.	Continuous limited frequent activity.	Frequent.	≤ 60% predicted; variability >30%

For intermittent → SOS β_2 -agonist

Mild persistent → +++ inhaled beclomethasone,
fluticasone, Budesonide.

Moderate persistent → +++ Salmeterol/Sustained release theophylline.

Severe persistent → ++ Oral low dose, long term, alternate day prednisolone.



2 yrs → Acute severe asthma
 ↓

Oxygen; Nebulise salbutamol
 ↓

Nebulised ipratropium bromide
 ↓

i.v. hydrocortisone
 ↓

s/c Terbutaline
 ↓

Terbutaline infusion
 ↓

50% $MgSO_4$ → aminophylline.

Foreign body:

CXR → Persistent inflation

Ball valve inflation.

Rx → Bronchoscopy & remove foreign body.

Recurrent pneumonia:

2 episodes of radiographic pneumonia in 1 year

(OR)

3 episodes in any time frame.

Persistent pneumonia:

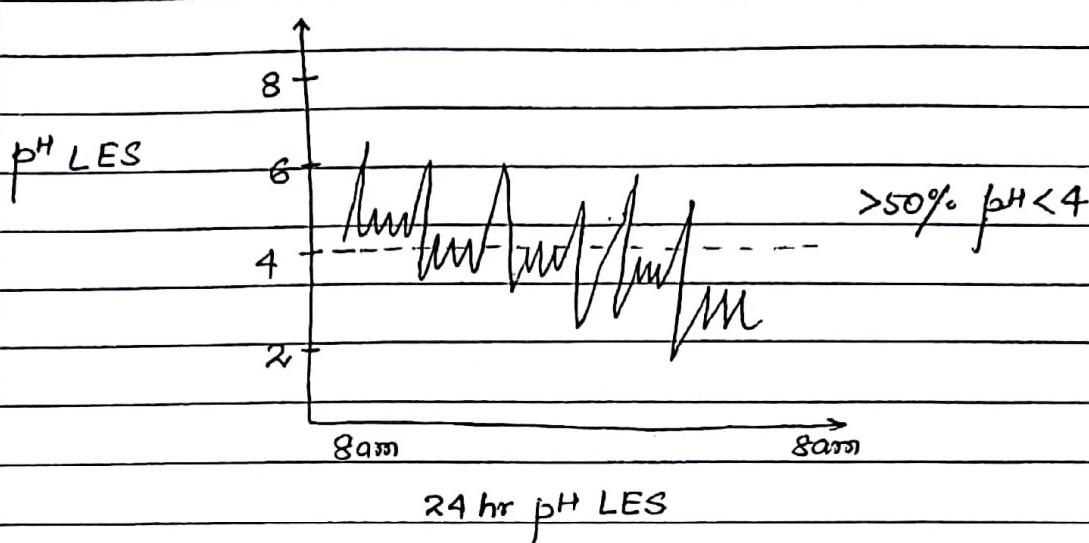
Persistence of symptoms & radiographic abnormalities for more than 1 month.

Recurrent pneumonia:

Cause → L → R shunt

- GERD dt aspiration.

- Immunodeficiency.



M/c/c of Recurrent pneumonia -in U.S. = Cystic Fibrosis.

Cystic Fibrosis

- Incidence: 1 in 2500 in UK

Gene → CFTR gene (7q 31.2 locus)



CAMP regulated chloride channel.

- In CF more Sodium goes to lumen ~~than~~ to mucus than chloride.

- Autosomal Recessive disorder.

- Respiratory: Pneumonia (>5 yrs of life).

M/c organism ass. c cystic fibrosis - *Pseudomonas aeruginosa*.

(Mucoid > Non Mucoid)

Rx:  Inhaler antibiotics

- Tobramycin
- Aztreonam
- Colistin
- Ciproflox

- Amikacin
- Levofloxacin

Respiratory : Pneumonia (>5 yrs)

- < 5 yrs - S. aureus; H. influenzae.
- 5-18 yrs - S. aureus; Pseudomonas
- > 18 yrs - Pseudomonas; S. aureus.

Acromonas; Acinetobacter;

Burkholderia cepacia

↳ specific; fatal

↳ Rx Mild - Colrimoxazole

↳ Severe - Meropenem +

Colrimoxazole/Doxycycline.

- Exocrine Pancreas - Steatorrhoea (< 5 yrs)
foul - bulky stools.



Rx - Steatorrhoea in CF

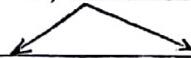
Lipase 1000 IU/kg supplement.

- Endocrine → 25% DM after 35 yrs.
- GIT → Meconium ileum.

Adolescents - Distal intestinal obstruction syndrome.

48hrs old baby has not passed meconium.

Ix → Lower GI contrast study



Diagnostic Therapeutic in meconium ileum.

- GIT: Diffuse pain abdomen
Colonic mucosal thickening.
Intussusceptions.

- Nasal polyps; Azoospermia

- Absent Vas : 1% infertile men ; common in CF males.

Q. Which glands are not obstructed in CF ?

- A) Cervix → Infertile
- B) Pancreas → Insufficiency.
- C) Sweat glands

→ causes "Traficking Defect"

M/c mutation in CF = ΔF508 mutation (Class II mutation)
(Seen in 70% caucasians)
↓ (25-30% Indian)

M/c lethal genetic disorder in caucasians.

CFTR gene has 1800 mutations.

How many Nucleotide deleted in CF ?

3 → TTT = Phenylalanine.

Rx : For Traficking Defect



DOC [LUMACAFTOR (Traficking corrector drug)

+

IVACAFTOR (Potentiator drug).



opens CFTR Cl⁻ channel.

Lumacaftor - induce hepatic Cyp 450



∴ New Corrector → TEZACAFTOR



doesn't induce Cyp 450.



Q. Which of the following is a calcineurin inhibitor?

- A) Tacrolimus
- B) Sirolimus → m-for inhibitor
- C) Everolimus]
- D) Cyclosporine

Diagnostic Criteria of CF:

- ① Sweat Chloride $> 60 \text{ meq/L}$ on 2 occasions
[$\text{N} < 40 \text{ meq/L}$]
- OR, ② Two known CFTR mutations.
- OR, Best - ③ Diagnostic nasal electrode potential difference.

Newborn screening for CF

IRT test \rightarrow Immuno reactive Trypsinogen
Assay test.
(Sensitive test)

Q. Male, 10 month boy, Down Syndrome & Recurrent pneumonia

Filling defect \rightarrow In Ba-swallow

\hookrightarrow in middle of esophagus.

\hookrightarrow Aberrant right subclavian artery
vascular ring



Dysphagia lusoria.

Approach to a pt. c recurrent / persistent pneumonia:

History, physical exam, CXR

↓
— Rule out TB

Difficulty in
feeding; choking
during feeds.

Infection in other
parts of body

Associated
Malabsorption.
Pseudomemos

No clues

GER studies.

Immunoglobulin

in airway

CD4, CD8

↓

Esophageal pH monitoring,
Barium Swallow.

NBT, HIV test.

Sweat Cl⁻ test

Direct laryngoscopy.

Mutation Studies.

Iso tonic fluids:

- 0.9% NaCl — 154 meq/L Na & Cl.

- Ringer lactate = Plasma

- 130 meq/L Na

- 109 meq/L Cl

- 28 meq/L Lactate

- 4 meq/L K

- 3 meq/L Ca

Maintenance fluid:

Need?

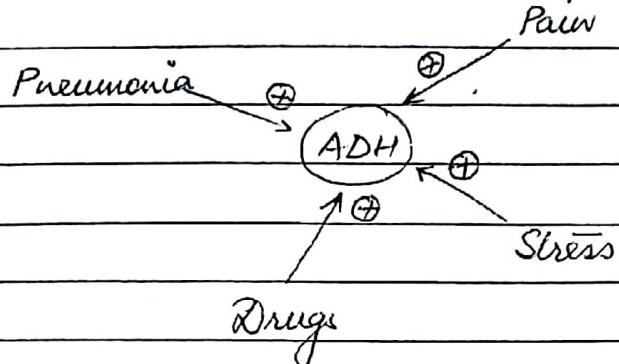
- Insensible water loss.

- Energy

- To prevent catabolism.

Type?

- Adult: 5% DNS
- Children: 5% Dextrose + N/2 or NS.

Amount?

A/C to Holiday & Segar

< 10 kg - 100 ml/kg/day.

10 kg - 1000 ml/day. (40 ml/hr).

11-20 kg → 1000 ml (for 10 kg) + 50 ml/kg additional kg above 10 kg.

> 20 kg → 1000 ml + 500 ml + 20 ml/kg additional above 20 kg.

Patau's Syndrome (Trisomy 13):

- Cleft lip & palate.
- Polydactyly
- Hypotelorism (Eye separated wide).

- Abnormal looks like Cyclops

↳ Holoprosencephaly.
(single eye)

↳ Fused frontal lobes + lateral ventricles.

↳ Aplasia cutis (problem of cleavage of skin).

~~- Rocker bottom~~

Edward Syndrome (Trisomy 18)

- Rocker bottom feet
- Overlapping of fingers.
- 2nd M/C Trisomy.
- Maternal age
- GIT anomaly are common
 - ↳ Atresia gut
 - ↳ Exomphalos
 - ↳ Malabsorption.

TTNB (Transient Tachypnoea of New Born) : Wet lungs.

R/F :

- Term ; Cesarean Section
- Macrosomia
- Precipitous labour.
- Maternal Sedation

CxR - Prominent horizontal fissure.

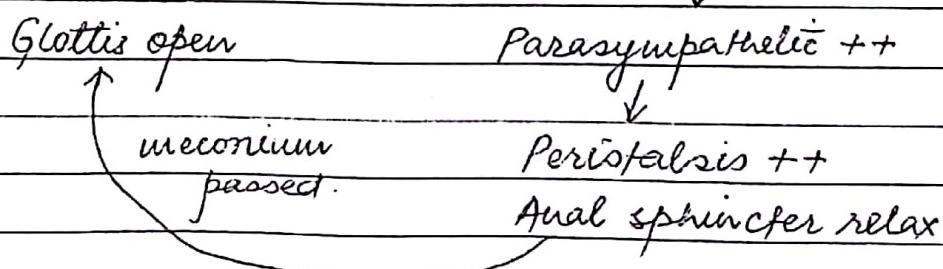
- It is a benign, self limiting condⁿ & resolves in 48-72 hrs.

FiO_2 requirement < 0.4

Never require mechanical ventilation.

MSL (Meconium Stained Liquor):

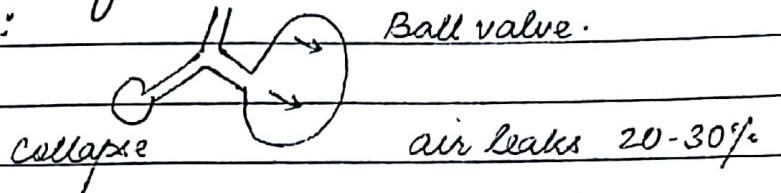
- Marker of perinatal hypoxia (common in Post term)





Complications of Meconium:

Physical :



Chemical : Irritant.

Biological \leftarrow impairs surfactant function (Surfactant _{given})
Good culture media (Given Antibiotic)

Rx : Meconium stained liquor



Born

Vigorous \leftarrow Zone
 HR > 100/min
 Resp effort.

YES

Transfer to mother

Liap or,
HR < 100 or,
Apnea.

NO
 \rightarrow
 PPV \times 100% O₂.

Transfer to mother

GROWTH

1-4 months : Weight gain 30gm/day

5-8 month : wt. gain @ 20gm/day

9-12 month : wt. gain @ 10gm/day.

Weight multiples:

wt. \times 2 = 5 months

\times 3 = 1 yr

\times 4 = 2 yrs

\times 5 = 3 yrs

\times 6 = 5 yrs

\times 7 = 7 yrs

\times 10 = 10 yrs.

Length:

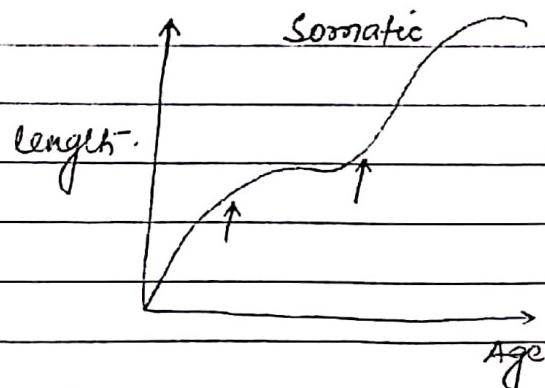
At birth - 50cm] Growth Velocity $\rightarrow +25$ (first year)

1 yr - 75 cm

2 yrs - 90 cm $\rightarrow +15 \rightarrow$ second year.

$4\frac{1}{2}$ yrs - 100cm

Add 6cm/year till puberty.

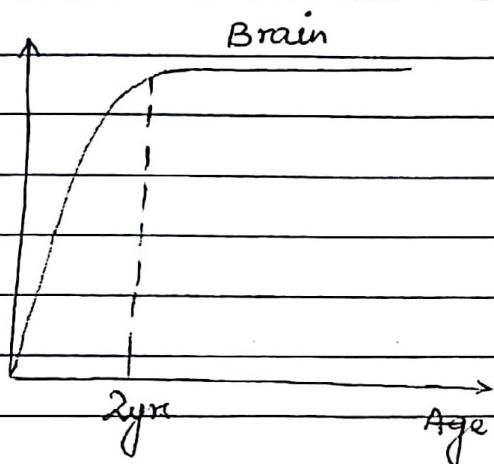


Q. In school going children; the avg height velocity is
 $\rightarrow 5-8$ cm

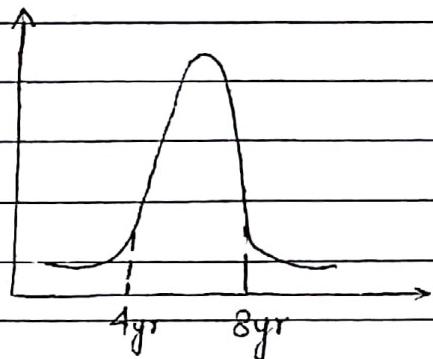


Head circumference:

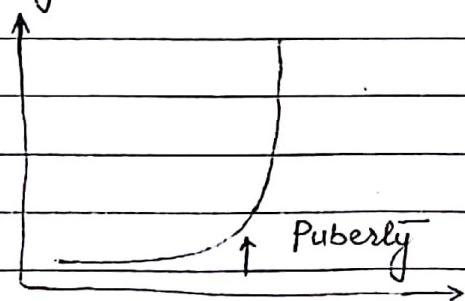
- 35cm at birth
- 3 months → 40cm
- 12 months → 45cm
- 2 yrs → 48cm (90% of brain grows)
- 12 yrs → 52cm.



Lymphoid growth:



Gonad growth:



Adolescent — 10 to 19 yrs.

Tanner's Sexual Maturity Rating:

SMR Stage I - V

Stage I — No character

Stage V — Completely developed.

Menarche → SMR - IV

Sequence of puberty:

Girls:

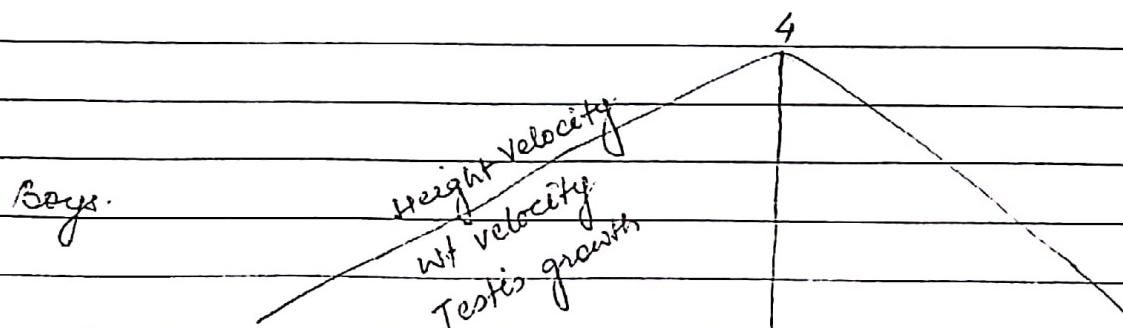
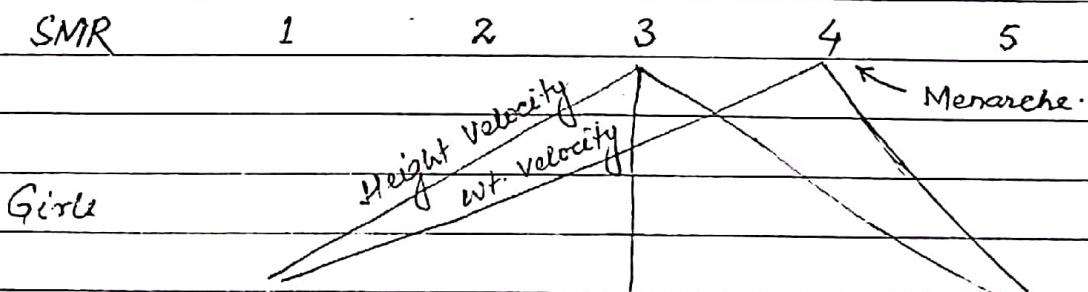
Growth Spurt → Thelarche, Pubarche, Menarche.

Boys:

→ Testis — Penis — Pubic hair, Axillary hair.

Girls: BP (Breast — Pubic hair)

Boys: GP (Genitalia — Pubic hair).



Most rapid ↑ in height → Phase 3 in girls & Phase 4 in boys.

Puberty - comes by pulsatile release of GnRH.

Precocious Puberty:

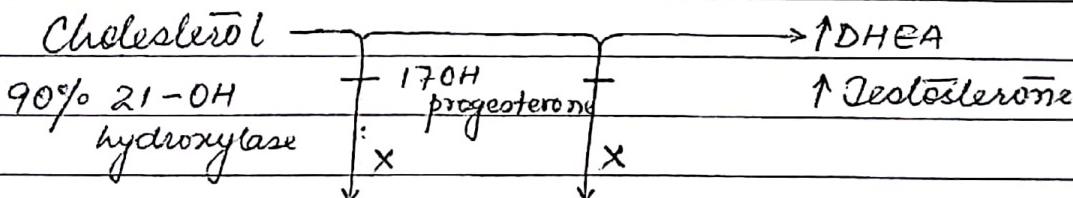
- Gonadotropin dependent / Central
- Females - Idiopathic.
- Males - Organic
(Hypothalamic Hamartoma,
Craniopharyngiomas,
Hydrocephalus, TBM).

CAH (Congenital Adrenal Hyperplasia):

M/c/c of female hermaphrodite.

Karyotype - XX

But looks like Male.



Cortisol	Aldosterone
glucose ↓	$\text{Na}^+ \downarrow$
Salt-Wasting Crisis (70%)	
$\uparrow \text{K}^+$	
Shock	

- Cause of precocious puberty in boys.

Genitalia → Ambiguous

Labia is pigmented d/t ACTH.

Virilization

Hypertrophy of clitoris

- Penoscrotal hypospadias
+ Empty Scrotum.

Due to deficiency of 21-OH hydroxylase lead to accumulation of 17-OH progesterone.

♀ Level of 17-OH progesterone in CAH

A) < 150

B) $150 - 300$

C) $300 - 500$

D) $> 600 (> 3500)$

CAH associated with Premature epiphyseal closure.



Short Stature.

Rx : CAH

- Supplement of Hydrocortisone & Fludrocortisone



Glucocorticoids

Mineralocorticoids.

$15-20 \text{ mg/m}^2/\text{day}$

0.15 mg/day .

Thrice daily.

- Girls require Sx

- Clitoroplasty

- Reconstruction Sx.

- Never get married.

Cholesterol

→ DHEA

Testosterone.

11β -OH
deficiency

11-deoxycortisol

↓
Cortisol

DOC: potent

mineralocorticoid

Aldosterone ↓
HTN

Hypokalemia metabolic
alkalosis.



Q. 5 yrs old boy has precocious puberty. BP = 130/80
Estimation of $\underline{\text{C}}$ help diagnosis?

- A) 17-OH progesterone
- B) 11-deoxy cortisol
- C) Aldosterone
- D) DOCA

Deficiency of 17 α -Hydroxylase \rightarrow \uparrow Aldosterone

\downarrow
HTN, Hypokalemia,
metabolic alkalosis.

\rightarrow X Cortisol
 \downarrow glucose.

\longrightarrow X Testosterone

\downarrow

Male look like ♀ .

(Mineralocorticoid) M

T (Testosterone).

α

\uparrow

\uparrow

\uparrow

\uparrow

\uparrow

3 β Hydroxysteroid dehydrogenase deficiency (3 β HSD def.)

\downarrow
Causes ambiguous genitalia in both sexes.

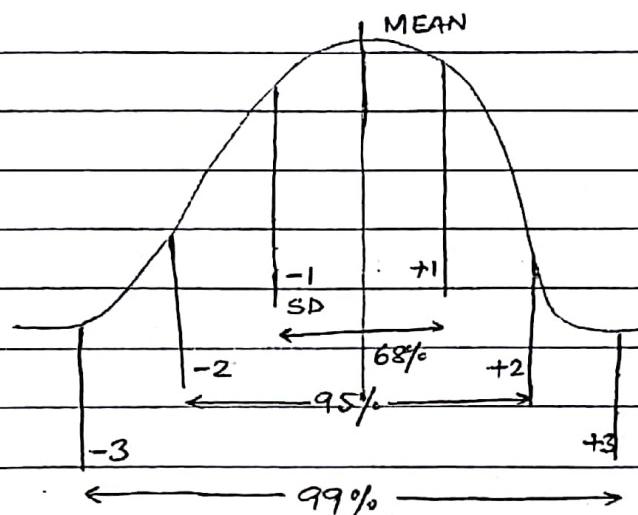
Q. Mother; previous child CAH

Next pregnancy - To prevent female virilization
 of foetus

Dexamethasone

(20mcg/kg pre pregnancy wt.)

Inhibits ACTH.



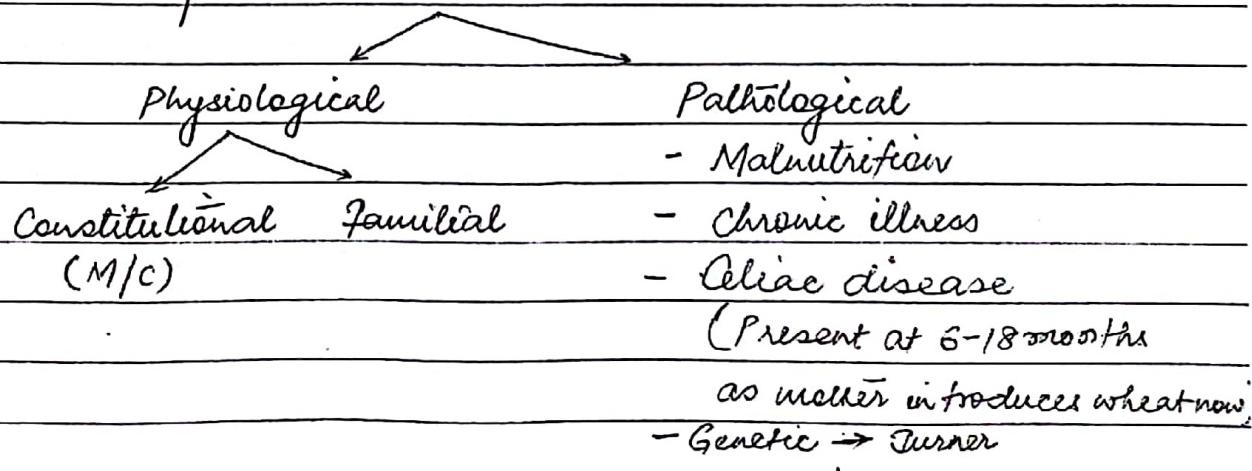
Short height > -2 SD below mean or < 3rd percentile

Mid parental height = Adult predicted height

Boys = average parents height + 6.5cm

Girls = " " - 6.5cm.

Causes of Short Stature:





Rx : Celiac ds

- Restrict wheat, Rye & Barley for lifetime.
- Gluten-free diet.
- Also avoid oats.



- Endocrinopathy → Hypothyroidism
Cushing
CAH
GH deficiency.

GH deficiency:

- Birth weight & length normal.
- Lag at 1-2 yrs of life.
- Bone age delayed.
- Doll face]
- Micropenis] Panhypopituitarism
- Short stature]

#. Micropenis ($\downarrow LH$, $\downarrow FSH$)

$\downarrow ACTH \rightarrow$ Hypoglycemia

- Hoarseness of voice.

INV: GH level $\geq IGF, BP3$

GH stimulation test - Best

\hookleftarrow Basal level GH

\hookleftarrow Stimulated level - 2 stimuli

(clonidine; insulin; L-arginine).

Rx - Recombinant GH (US, FDA)

Indication: rGH

- GH deficiency
- Turner Syndrome
- Prader willi
- Chronic kidney ds.
- SGA height - 2.25 SD below mean

When to stop GH?

- Height reaches 50th centile
- Epiphysis fuses.
- Pseudotumor cerebri
- Slipped capital femoral epiphysis.

Disproportionate short stature:

US/LS ↑ → Achondroplasia, Rickets;

Hypothyroidism.

US/LS ↓ → TB spine, Mucopolysaccharoidosis IV

(Morgni's ds)

Physiological Short Stature:

	CONSTITUTIONAL (M/c)	FAMILIAL
Birth length	• (N)	IUGR
Lag	6-12 months	
Growth velocity	(N)	Less
Final Height	(N) / Sub (N)	Less
Puberty	Delayed	(N) \neq
Bone age	Delayed.	(N) \neq



Developmental milestones:

Neck holding → @ 3 month.

Gross motor milestones :

3 months - Head holding; Neck holding.

5 months - Sitting \pm support.

4-6 months - Prone to supine, supine to prone
(Roll in bed).

8 months - Sitting \pm out support.

9 months - Crawling

10 " - Creeping; stand \pm support

12 " - Standing \pm out support; walking \pm out support.

2 yr - Walk up stairs \pm two feet at each step.

3 yr - Upstairs \pm one foot at each step, rides tricycle.

4 yr - Hop on one foot

5 yr - Skips on two foot

Fine motor :

12 wks \rightarrow Moro's reflex disappears

Grasp reflex disappears

4 months - Goes for objects

5 months - Bidextrous grasp.

6-7 " - Transfer object, palmar grasp.

9 " - Pincher grasp; mature meat.

13 months - Casting.

15 " - Self feed \pm a spoon

18 " - Self feed \pm a cup.

24 " - turns pages of a book one at time.

Social Milestone:

2 months - Social smile

3 " - Recognizes mother.

6-7 " - Smiles at mirror image.

9 months - Waves bye-bye.

6 months - Stranger anxiety.

2 yrs - Dry by day

3 yrs - Dry by night

Dress/ undress himself (Supervision)



Out supervision - 5 yrs.

18 months - Separation anxiety/ Clinginess/ Reproachment.

Language Milestones:

1 month → Head turns to sound

3 months → Cooing

6 months → Babbles; Monosyllables (ma, ba)

9 months → Bisyllables (mama; ba-ba)

1 yr → 2 words & meaning

18 months → Vocabulary of ten words.

2 yrs → Simple sentences & 2 words; Phrase.

3 yrs → tells age & sex; uses pronouns,

handedness; identify colours.

4 yrs → tells story

5 yrs → Knows colour

15 months : 4-5 words

2 yrs → 50-100 words

When can a child understand death =

< 3 yrs - No idea

3-9 yrs - Idea

> 9 yrs - Entity ; Irreversible ; himself

Object permanence / Constancy - 9 months.

Cross a busy road - 10-12 yrs.

Tie shoe lace - 5 yrs.

Bladder control - 85% by 5 yrs.

Nocturnal enuresis - > 5 yrs.

↳ more than 2/week x 3 months.

Q. 14 yrs old child c Nocturnal enuresis

Rx - • Behavioural - +ve reinforcement.*

• Alarm therapy (Best)

↳ 85% relapse free rates.

• Drugs → Desmopressin

Encopresis : > 4 yrs

Cause → Chronic constipation.



CNS

MICROCEPHALY:

$\rightarrow HC > -3 SD$ below mean

Causes: GENETIC

- Trisomy 13, 18, 21
- Cri-du-chat Syndrome

ACQUIRED

- Baby: HIE; Hypoglycaemia, PKU, Meningitis, encephalitis.
- Mother \rightarrow TORCH, ^{lalanine} Hyperphenylalanine $> 6 \text{ mg/dl}$
Alcohol, DM, Radiation.

MACROCEPHALY

$HC > 2 SD$ above mean

Q Neurodegenerative disorders:

GREY MATTER:

- Normal at birth.
- Regression milestones.
 \hookrightarrow Disappear as they come orderly.
- Deaf, blind, seizures.
- Anemia, Hepatosplenomegaly.
- Cherry red spot macula.

Microcephaly

- GM1 gangliosidosis.

- Gaucher's ds

(β -gluco

- Niemann Pick's ds

Huge
spleen

Macrocephaly

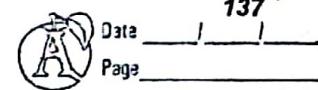
- GM2 gangliosidosis

(Tay Sachs' ds)

- Sandhoff

(

VLCFA = Very long chain fatty acid.



Tay Sachs ds -

- AR
- D/t deficiency of β -hexoseaminidase A.
- 1/25 Ashkenazi Jew are carrier.
- 6 months exaggerated startle reflex.
- Cherry red spot macule
- Organomegaly not seen.

Organomegaly + Tay Sachs = Sandhoff



Def. of β -hexoseaminidase A & B.

WHITE MATTER DISORDERS:

- All the tracts are white matter.
- Frequent fall, incoordination.
- Upper motor neuron sign.

Microcephaly / N

- Krabbe's
- XLR adrenoleukodystrophy.

↓
degeneration starts from
parieto-occipetal area.

△ → ↑ VLCFA levels

Rx → Early bone marrow Tx.

Lorenzo oil

- Metachromatid

leukodystrophy

Macrocephaly

- Canavan ds.

Alexander ds.

→ MRI → Diffuse white matter thickening.

→ MRI → Degeneration starts from
frontal periventricular area.



Hydrocephalus:

- Enlarged ventricles & or & out ↑ in ICT.

CSF production:

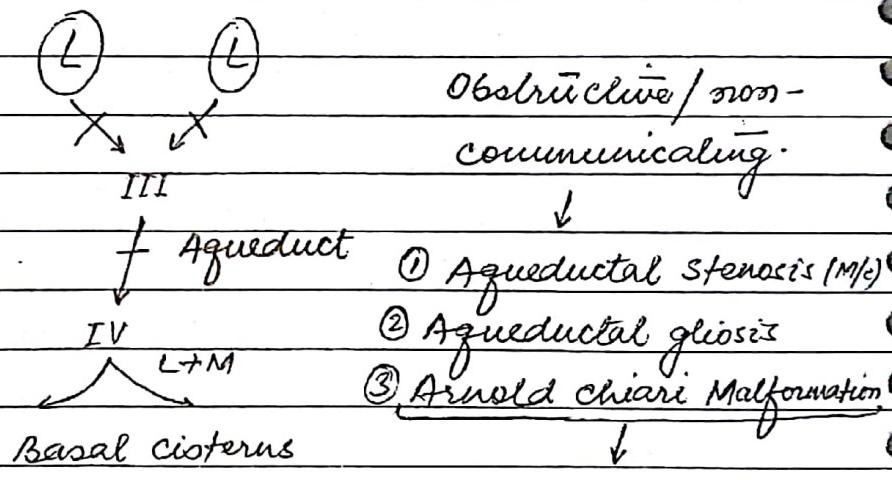
- Choroid plexus (75%) → lateral, III & IV ventricles.
- Extrachoroidal (25%) → Capillary endothelium.
in brain parenchyma.

Rate of CSF production → 20 ml/hr.

CSF volume in infants = 50 ml

adults = 150 ml.

CSF flow:



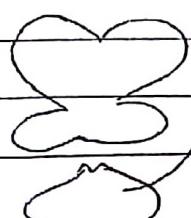
↓
Downward displacement
& hypoplasia in cerebellum
leading to obliteration of
cisterna magna.

Arnold - Chiari Type I - Adolescent/Adult

Type II - Newborn &

lumbosacral myelomeningocele

④ Dandy - Walker Syndrome:



→ large cyst in posterior fossa communicating
& 4th ventricle.

- child has cerebellar hypoplasia

⑤ Vein of Galen malformation:

- M/C arteriovenous malformation in brain
- Sinus Venosus ASD.
- Obstructs aqueduct.
- Midline mass & dilated lateral ventricle.

Non-obstructive / communicating :

- Basal exudates

↳ TB

↳ Cryptococcal meningitis.

IOC for congenital Hydrocephalus — MRI.

Rx: Drugs → Furosemide
 Aclazolamide.

VP shunts → anastomose ~~to~~ ventricles to peritoneum.

↳ Gross hydrocephalus

↳ Deviated parenchyma.

↳ Complication — Blockage

Infection (Coagulase
-ve staph).



Febrile Seizures:

- M/c seizure during childhood.
- Between 6 months - 5 yrs.

R/F for recurrence:

Major:

Age < 1 yr

Duration of fever < 24 hr.

Fever 38-39°C (100.4-102.2°F)

Minor:

Family H/o Febrile seizures.

Family H/o epilepsy.

Complex febrile seizures.

Daycare

Male gender.

Lower Serum Na⁺ at time of presentation.

Having no risk factors carries a recurrence risk of approx 12%

if 1 R/F → 25-50%

2 " → 50-59%

≥ 3 " → 73-100%.

R/F	Risk for Subsequent Epilepsy
Simple febrile Seizure	1%
Recurrent "	4%
Complex "	6%
(> 15 min duration or recurrent ≥ in 24 hrs)	
Fever < 1 before febrile seizure	11%
Family H/o epilepsy	18%
Neurodevelopmental abnormality (Mental Retardation).	33%

Complex febrile seizure complication:

- Prolonged febrile seizures.
MRI → Mesial temporal sclerosis

Temporal lobe + Hippocampal lobe.

Epilepsy in children:

Partial:

- Simple
- Complex - aura; automatisms.

Rx — Oxcarbamazepine &
Carbamazepine.

Ring Enhancing lesions.

Neurocysticercosis

- Solitary
- SCOLEX

Rx — Albendazole (DOC)

↓

Before giving Albendazole

3-5 days of steroids given.

Tuberculoma.

- Large $> 20\text{mm}$
- Multiple
- Irregular margin
- Perilesional edema

— Midline shift.

Generalised epilepsy:

- Tonic
- Clonic
- GTCS [aura - GTCS → postictal phase]

↓

Drowsy, unconscious,
frothing, tongue bite
uprolling, incontinence.

- Atonic
- Myotonic



Rx - Sodium Valproate.

↓
In < 2 yrs → It is hepatotoxic.

Absence Seizure:

- Blank stare < 30 sec.
- No aura/post ictal phase.
- Hyperventilation provokes.

Rx : Ethosuximide (DOC)
Valproate

Atypical Absence seizure:

- Myoclonic component

Rx - Valproate.

JME (Juvenile Myoclonic epilepsy):

- 12-18 yrs.
- Myoclonic jerks morning.
- Drops things.
- Gene by gene
- Family History.
- GTCS seizures 90%
- 1/3 Absence seizures.

EEG of JME → Generalised 4-6 Hz spike
+ photic stimulation.

Rx - Valproate (lifelong) → Excellent.

Infantile spasms / Salaam / West

- Flexor contractions of Head, trunk & extremities.

- 4-8 months

- EEG → HYP SARRHYTHMIA

↳ Generalised chaotic high volume slow wave.

- Idiopathic / Cryptogenic → Good.

- Secondary → HIE, Structural malformation, Down's Syndrome, Tuberous sclerosis.

Rx: — ACTH → inhibit CRH. (DOC).

Vigabatrin → In Tuberous Sclerosis

STATUS EPILEPTICUS:

- Convulsion $>$ 30 min

(OK)

Continuous b/w no regain of consciousness.

Vulnerable to hypoxia — Hippocampus, amygdala, Thalamus, subcortical areas.

Rx: - i.v. Lorazepam (DOC) — longer t_{1/2}
0.05 mg/kg

- Midazolam + Phenytoin (20mg/kg)

Repeat 10mg/kg — 10mg/kg

Phenobarbitone 20 mg/kg — 10 mg/kg

i.v. Valproate 20-30 mg/kg

Midazolam infusion 2-20 mcg/kg/min.

i.v. ~~Lamotrigine~~ Levetiracetam 20 mg/kg.

GA propofol; Thiopentone.



M/c/c of Status epilepticus → Febrile Seizures.

↓
Rx → per rectal diazepam /
Buccal midazolam

Prevention of Febrile Seizures:

- No need

- Risk of Recurrence / concerned parents



Intermittent prophylaxis Oral CLOBAZAM / DIAZEPAM

↳ New BZD

for 48-72 hrs of fever.

MENINGITIS:

Cause :

	India	World
< 2 months	Klebsiella E. coli	Gr.B / D Streptococci E. coli
2 months - 3 yrs	H. influenzae type B	S. pneumoniae.
> 3 yrs	S. pneumoniae	S. pneumoniae Nisseria

Acute Bacterial meningitis:

- 95% cases occurs b/w 1 month to 5 yrs.
- Defect of complement system C₅-C₈ & properdin system
 - meningococcal infection
- T-lymphocyte defects (eg - AIDS / chemotherapy)
 - Listeria monocytogenes / cryptococcus.

- congenital/acquired defects across mucocutaneous barrier → Pneumococci d/t cribiform plate.
- lumbosacral meningo myelocoele & dermal sinus - staph. & enteric bacteria.
- Penetrating CNS trauma / CSF shunt infection
 - coagulase - ve staph.

Recurrent meningitis in CSF leak pt. M/c d/t β pneumococcus.

Splenectomy vaccination time - 2 wks before.

Rx: Ceftriaxone (D/C)

~~20% of resistant pneumococci = Vancomycin~~

+ Ceftriaxone.

M/c Neurological sequel of meningitis:

- SNHL via aqueduct cochlear.

- Can we prevent SNHL?

- Dexamethasone

- 0.15 mg/kg

- 30-60 mins before antibiotics.

Post exposure prophylaxis to contacts & doctors -

H. influenzae & *Nisseria* → imm. Single dose ceftriaxone.

- Rifampicin X 2 days

~~Doctors~~ → Fluorquinolones.



Q. 3 yrs old diagnosed to have HiB meningitis. Tx done before discharge → BERA.
(Brainstem evoked response audiometry)

ENCEPHALITIS:

M/c/c = Enteroviruses (80% cases).

M/c sporadic = HSV-1

M/c aseptic meningitis in unimmunized children
— mumps.

Q. Child c fever & coma

Focal seizures.

CSF: Hemorrhagic

CT: Temporal hypodense ; MRI → Hyperintense
— HSV-1 infection.

Localised temporal spike → HSV encephalitis.



DOC: i.v. Acyclovir.

Mortality rate of untreated herpes = 70%.

AFP (Acute Flaccid paralysis):

- Acute onset < 6 weeks

- < 15 yrs

- Rule out pseudoparalysis

↳ Septic arthritis

Osteomyelitis

Scurvy; early syphilis

Hypokalemia → Hypotonia.

- Asymmetrical AFP:

- Paralytic polio.

• Traumatic neuritis → i.m. injection (d/t)

- Symmetrical AFP:

• Transverse myelitis

↳ Herpes, varicella, mycoplasma

Level → Thoracic area.

Rx: high dose i.v. Methyl prednisolone.

• Guillain Barre Syndrome (ATDP)

↳ Demyelinating.

- Diarrhea (By Campylobacter jejuni).

- Weakness occurs after 10 days.

- Areflexia (DTR absent)

- Symmetric

- Ascending → diaphragm involved.

- Plateaus → 4 wks.

- Sensory & autonomic changes.

- Also d/t Mycoplasma; salmonella;

S. pneumoniae.

CSF	1st wk	2nd wk
Cells	10/lpf	10
Proteins	50 mg/dl	500

→ Albuminocytological dissociation

- B/L symmetric demyelinating illness.

IOC for calcification: CT scan.

Rx: i.v. Ig (2g/kg)

↓ fails

Plasmapheresis.



Iv Ig indications:

- ↳ Kawasaki
- ↳ ALD P (Acute inflammatory Demyelinating Polyradiculo neuropathy)
- ↳ Hypogammaglobulinemia
- ↳ Rh isoimmunisation.

General Pediatrics:

- Diarrhea
- ORS
- Zinc
- Malnutrition:

ORS:

Residual Mmol/L		Old (WHO-ORS)	New / Universal
45	Sodium	90 (Cholera stool loss)	75 (Rotavirus 50-70)
40	Potassium	20	20
125	Glucose	111	75 (for facilitate diffusion of Na ⁺ in cell)
70	Chloride	80	65
7	Citrate	10	10
+Mn,Zn,Cu		311	245
300			low osmolarity ORS.

Citrate improves the shelf life of ORS.

Resomal → Rehydration solⁿ for malnutrition.

Hyperkalemia Hypernatremia.

Substrate concentration of components of ORS solⁿ:

NaCl — 2.6 gms

KCl — 1.5 gms

Trisodium citrate — 2.9 gm

Glucose, anhydrous — 13.5 gm

WHO dehydration:

- No
- Some → Skin pinch slowly; thirsty
- Severe → skin pinch very slowly; lethargic; oliguria.

Rx: No dehydration:

- Replace ongoing losses.
- 5-10 ml/kg.

Some dehydration:

- ORS 75 ml/kg over 4 hrs.

Severe dehydration:

- i.v. Ringer lactate 100 ml/kg.

	30 ml/kg	70 ml/kg
>1 yr	0.5 hr	2.5 hrs
<1 yr	1 hr	5 hrs.

WHO Zn in Acute Diarrhea:

- 2X RDA for 2 weeks.

Zn dose	RDA	Diarrhea
>6 months	10 mg	20 mg.
<6 months	5 mg	10 mg

Aerodermatitis enteropathica (periorificial dermatitis)

- d/t Zn deficiency.
- Nutritional
- Genetic (AR) → Intestinal Zn transporter defect.
- Low Zn levels.

- Improve on Zn supplement.

Rx - Zn (3mg/kg) — elemental Zn.

Malnutrition:

	Marasmus	Kwashiorkar
Muscle Wasting	++	++
Edema		++
Hepatomegaly		++
Low albumin		++
Pigment		++
Aptite	Voracious	Poor
Sensorium	Alert	lethargy.

Weight/Age criteria (Indian Academy of Pediatrics)

- Normal > 80% reference.
- Grade I → 71 - 80%
- Grade II → 61 - 70%
- Grade III → 51 - 60%
- Grade IV → ≤ 50%

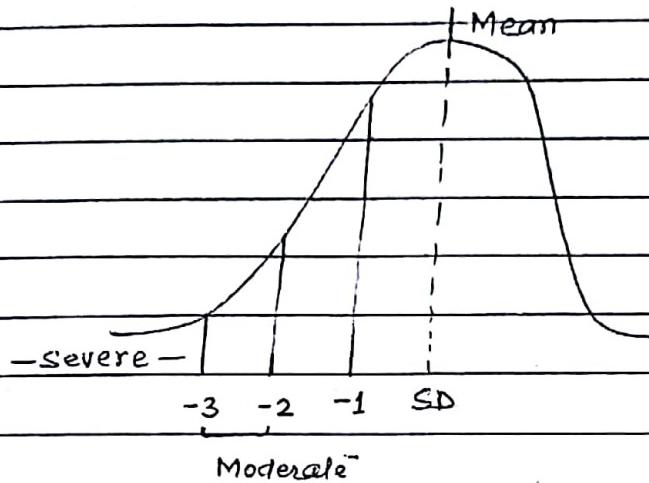
for Edema ⇒ Add K
 ↳ Bad marker.

WHO

	Moderate	Severe
Weight/Height	71 - 79%	≤ 70
Acute (Wasting)		
(N) > 80% reference		
Height/age		
Chronic (Stunting)	86 - 89%	< 85%
(N) > 90% reference		

Symmetrical Edema

+++



Severe Acute Malnutrition:

Among children - 6-59 months of age.

Any of the following:

- ① Weight for height below -3 standard deviation (SD or Z scores) of the median WHO growth references.
- ② Visible severe wasting.
- ③ Presence of bidental edema.
- ④ Mid-arm circumference below 11.5 cm.

Below 6 months → Mid-arm circumference can't be used.

Criteria for admission - If child fails appetite test.

Criteria for passing Appetite test.

Body wt. (kg) Min^{mm} amount of RUTF (Ready to use therapeutic factor) to be consumed for passing Appetite test (ml or grams)

< 4 kg

15 mL

4-6.9

25 mL



7 - 9.9 35 mL
10 - 14.9 50 mL

Mx :

Complications of SAM

Mx C in S = Sugar (Blood Sugar < 54 mg/dL OR < 3 mmol/L)

1st 2 day L H = Hypothermia (< 95.5°F OR < 35.5°C)

C in 1st wk I = Infection (TB, Malaria, UTI)

EL = Electrolyte (Hypokalemia & Hypernatremia)

DE = Dehydration

D = Deficiency of Vitamins & minerals

PHASE

STABILIZATION

REHABILITATION

Step

Day 1-2

Days 3-7

Weeks 2-6

1. Hypoglycemia

→

2. Hypothermia

→

3. Dehydration

→

4. Electrolytes

→

5. Infection

→

6. Micronutrients

No iron

iron

7. Cautious feeding

→

8. Catch up growth.

→

9. Sensory stimulation

→

10. Prepare for follow up.

→

Feeding Rehabilitation: Cautious feeding

- F-75 containing 75 kcal/100mL & 0.9 g protein/100mL

B = Begin feeds

E = Energy dense feeds

S = Stimulation

T = Tender love & care

Days	Frequency	Vol/kg/feed	Vol/kg/day
1-2	2 hrly	11 ml	130 ml
3-5	3 hrly	16 ml	130 ml
6-7	4 hrly	22 ml	130 ml

Energy dense feeds:

After the transition give:

- Frequent feeds (at least 4-hrly) of unlimited amounts of a catch-up formula.
- 150 - 220 kcal/kg/day.
- 4-6 gm protein/kg/day.

Criteria for discharge:

- Weight for height > 80% of Reference standard.
- Edema should be absent for 2 wks.
- MAC > 12.5 cm.
- Weight gain > 5g/kg/day X 3 days.
- Appetite is good.
- Complete antibiotics.
- Care taker should have learnt, motivated.

Weight for age < Acute (wasting)
Chronic (stunting)

Age independent criteria — MAC (>12.5cm)

b/w 1 to 5 yrs.

Age independent index — Kanawati & McLaren index

Rao & Singh index

Dugdale index.

Quac stick index

Telliffe ratio



$$\# \text{ Osmolality} = 2 \times [\text{Na}] + [\text{Glucose}] / 18 + [\text{BUN}] / 2.8.$$

Pediatric Nephrology:

- Development
 - Oliguria; polyuria
 - Hematuria
 - AKI
 - CKD
 - Nephrotic, Nephritic
 - UTI
- Topic

Development:

GFR:

- Newborn $\rightarrow 15 - 20 \text{ m}\text{l/min}/1.73 \text{ m}^2$
- 3 months $\rightarrow 2/3^{\text{rd}}$ adult
- Like adults $\rightarrow 2 \text{ yrs of life.}$

- Tubular concⁿ:

- Adult morning urine osmolality $> 800 \text{ mosm/kg.}$
- like adult 1 yr.

- Nephrogenesis complete @ 36 wk of gestation.

- Barker's hypothesis:

preterm & IUGR \rightarrow hypertension in 2nd to 3rd decade
 ↳ Bcoz they have less no. of nephron.

- M/c asymptomatic abd. mass in 1-5yr - Wilms tumour

- M/c abd. mass in newborn - Multicystic

dysplastic kidney.
 ↓

non-functional mass \rightarrow bunch of grapes

- 80% U/L.

OLIGURIA:

- U.O. $< 1 \text{ ml/kg/hr}$
- Common in AKI & Acute GN.

Causes of Non oliguric AKI — Aminoglycosides

- Neonatal renal failure.
- Resolving ATN (polyuria)

POLYURIA:

- U.O. $> 4-5 \text{ ml/kg/hr}$. [Polydipsia; Polyuria]

Approach —

- Blood glucose [RBS $> 200 \text{ mg/dl}$
or FBS $> 126 \text{ mg/dl}$]

↳ DM

- Venous Blood Gas.

(i) Hypokalemic hypochloric metabolic alkalosis



Bartter Syndrome; Gitelman Syndrome.

Bartter Syndrome

- Severe; Early.
- Infancy
- Antenatal \rightarrow Polyhydramnios
- Hypercalcemia

Gitelman Syndrome

- Mild; Well preserved.
- Older child
- HypoMg
- Hypocalciuria.

(ii) Hyper Cl, hyper K, Normal anion gap
metabolic acidosis



Renal tubular acidosis.

(iii) Normal \leftarrow Psychogenic \rightarrow water deprivation

DI

\rightarrow in psychogenic →
Urine osmolality doubles



$\text{DI} \rightarrow \text{Central/XLR Nephrogenic}$

- On Vasopressin challenge, urine osmolality doubles by 100% in central. Hence differentiated from XLR Nephrogenic.

$\text{Q 4yr; Polyuria \& Polydipsia}$

Venous blood gas normal

Urine Osmolality

- Baseline 50 mosm/kg.
- On water deprivation = 60 mosm/kg
- On Vasopressin challenge = 70 mosm/kg.

$\Delta = \text{XLR Nephrogenic DI}$

DOC = Thiazides.

Best method of GFR estimation = Insulin clearance.

Schwartz eGFR formula:

$$= K \times \frac{\text{height in cm}}{\text{creatinine (mg/dl)}}$$

Formula depends on:

- Height, muscle mass.
- Method of estimation of creatinine.
[Jaffei reaction].

S. creatinine is accurately measured by

- Enzyme assay / HPLC.

Value of k in Schwartz formula:

Low birth weight infant — 0.33

Normal infant 0-18 months — 0.45

Girls 2-16 yrs — 0.55

Boys 2-13 yrs — 0.55

Boys 13-16 yrs — 0.70

Schwartz method is independent of — Renal ~~function~~
~~function~~

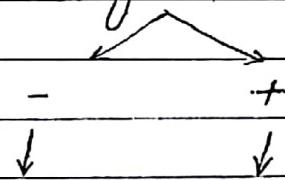
HEMATURIA:

Red urine:

- Beet root
- Phenolphthalein
- Rifaupicin

Q. Syms old, Red urine. Urine 1-2 RBC's/hpf —
Intravascular hemolysis.

If Red urine → Look for RBC's.



Hemoglobinuria Hematuria
Myoglobinuria.

Hematuria:

- Gross

- Microscope → > 5 RBCs/hpf on centrifuged urine



Cause —	Glomerular	Extrā glomerular
	- IgA ; MPGN; PSGN.	- Cystitis; Stones;

Idiopathic hypercalciuria
↓

24 hr urine $\text{Ca}^{2+} > 4 \text{ mg/kg}$.

Dysmorphic RBC ++

Colour Color Bright Red.

Pain Painless Painful.

Protein in urine Proteinuria

Recurrent Gross Hematuria:

- IgA
- MPGN
- Idiopathic hypercalciuria.

PSGN doesn't reoccur.

Alopecia Syndrome:

Traid :

Eye : Ant. Leucocoria

Ear : SNHL

Kidney : 75% Boys ESRD before < 30 yrs.

- 80% X linked > AR 15% > AD 5%
- Collagen IV → α -5 domain (in GB memb)

Good pasture Syndrome — d 3 domain of collagen IV abnormality.

INV: Electron Microscopy.

↳ Basement membr' is irregular.

↳ Splitting of the lamina densa

↳ Lamellation

↳ Striated GBM

↳ Basket weave appearance (classical)

Slit lamp exam → Keratoconus

Lenticconus

Acute Kidney Injury (AKI):

Best Biomarker → Urine NGAL

[Neutrophilic gelatinase associated lipocalcin].

② Urine IL-18

③ Urine KIM-1 (Kidney injury molecule)

④ Urine L-FABP (Fatty acid binding protein).

⑤ Serum cystatin-C

Types:

- Prerenal

- Renal

- Post renal

PRERENAL - Hypotension, Hypoxia

Newborns; hypovolemia, burns, diarrhea.

RENAL - ATN > HUS

POST RENAL - Obstruction.



Indices	Pre renal	-Renal
Urinary Sodium (meq/L)	< 20	> 40
Urine Osmolality	> 500	< 300
B-urea / Creatinine ratio	> 20 : 1	< 20 : 1
Fractional excretion of Na%	< 1	> 1
= $\frac{\text{Urine Na}}{\text{Serum Na}} \times \frac{\text{Serum Cr}}{\text{Urine Cr}}$		

Cause of AKI:

M/c/c in children/Adults

Prerenal → ATN

↳ Hypovolaemia

Hypotension

Drugs ↳ Exo ↳ Toxins

Sepsis ↴

Hemoglobinuria ↳ Endo ↴

Myoglobinuria

Hemolytic Uremic Syndrome (HUS):

- Microangiopathic hemolytic anaemia
- Thrombocytopenia
- AKI

• 90% follows diarrhea.

• Developed : E. coli O157:H7.

• Developing country : Shigella dysenteriae type I.

• Germany June 2011 : E. coli O104:H4

SHiga toxin → Cause Endothelial injury



TMA (Thrombotic microangiopathy).

On PBS → Schistocytes are classic of HUS

Rx : ECULIZUMAB [DOC] → for PNH

↳ drug against Cs.

If not available → Plasmapheresis.

Complications :

→ Insensible losses - 400ml/m²

- Fluid overload → Rx → Fluid restriction
- Hyperkalemia → cause arrhythmia / Sudden death.
- Dilutional hypo Na. Rx - Restrict fluid
- Dilutional anaemia Rx - PRBC if Hb < 6
- Metabolic acidosis.
- Hyper PO₄ → Hypo Ca
- HTN.

Hyperkalemia :

Rx - Glucose & insulin intravenously



C in 10-15 min.

- Ca²⁺ stabilises cardiac membrane potential.

↳ only given in ECG changes in Hyperkalemia.



Calcium Gluconate i.v.

~~Mechanism~~: Hyperkalemia Rx :

Transcellular shift into cell.

- Insulin & Dextrose.

- Nebulised salbutamol.

Cardioprotective → i.v. Calcium Gluconate.

↑ delivery Na to distal

• Furosemide

• i.v. NaHCO₃

Dialysis . . .



K-binders → Kaexylate polystyrene.

CKD (Chronic kidney disease):

Causes in children -

- < 5 yrs - Hypoplasia
Dysplasia
Posterior urethral valves (Boys)
- > 5 yrs - Acquired
↳ GN/HUS.

Features:

A = Azotemia

Acidosis (metabolic)

Anemia (Normocytic Normochromic)

↳ Rx - S/C rh EPO (Erythropoietin).

B = Bone ds

C = Cardiovascular complication ↑

G = Growth failure ← Multifactorial causes.

When GFR ↓ - 40-60 ml/min/1.73m²



↑ PO₄

↓ 1d-OH

↑ PTH

↓

↓ 1,25 vit.D.

↓ Ca⁺⁺

Osteodystrophy
(CKD - MBD)

GFR

Hyperfiltration injury

ESRD < 10 ml/min/1.73m²

Age

Nephrotic Syndrome:

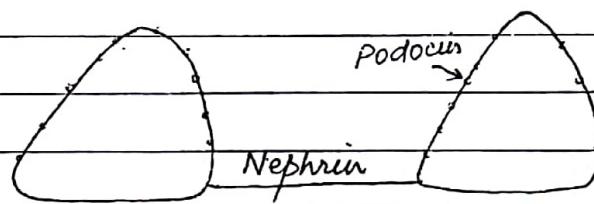
- Proteinuria $> 40 \text{ mg/m}^2/\text{hr}$ or $> 2 \text{ g}/24\text{hr}$
- Hypoproteinemia (Hypoalbuminemia)
- Hyperlipidemia (Cholesterol $> 200 \text{ mg/dL}$)
- Edema: S. Albumin $< 2.5 \text{ g/dL}$.

On light microscopy → Minimal changes.

On Electron " → Effacement of foot processes of podocytes.

DOC: Prednisolone

Cause of Edema in NS → Na^+ & H_2O reabsorption.



Gene	Protein	Disease
NPHS1 (Chr 19)	Nephrin	Finnish congenital nephrotic (< 3 months)
		Rx: Nephrectomy.
NPHS2 (Chr 1)	Podocin	Steroid resistant FSGS Rx: Catecholamine inhibitors ↳ Tacrolimus > Cyclosporine

Steroid toxic; Steroid dependent

- Cushingoid
- HTN
- Post-subcapular cataract
- Impaired glucose tolerance
- Short

Rx - Oral cyclophosphamide for 12 weeks.



UTI (Urinary Tract Infection):

Definition: Symptom + Urine culture $> 10^5$ CFU/ml

- Symptom - Fever
- E. coli
- Females [Boys < 1 yr]
- Ascending [$< 1 \text{ yr} \rightarrow \text{Hematogenous}$]

Best urine specimen - Suprapubic aspiration.

- Asymptomatic bacteriuria \rightarrow Shouldn't be treated.
- M/c/c of UTI in children \rightarrow VUR
IOC for VUR \rightarrow MCU

VUR :

- Polar scanning \rightarrow DMSA nuclear scan
- Function \rightarrow MAG3 / DTPA nuclear scan.
- Prophylaxis \rightarrow Antibiotic of choice

\downarrow
Cotrimoxazole.

Sx \rightarrow Reimplantation of both ureter.

\hookrightarrow Indication:

- Breakthrough UTIs
- Deterioration of Renal function.

GLOMERULONEPHRITIS:

- Hematuria
- Edema
- HTN

M/C/C of GN → Post streptococcal GN.

Acute post streptococcal GN:

- Follows infection of throat (serotype 12) / skin (serotype 49) C nephrogenic strains of β -hemolytic streptococci.
- Age \rightarrow 5-12 yrs
- Acute phase resolves in 4-6 wks but urine normalises in 1 yr.

4-6 wks

Edema

X

Cala Gross hematuria

X

HTN

X

C_3

6-8 wks
normalise.

In urine - Shows microscopic hematuria

6-12 months to resolve.

Sore throat $\rightarrow \Delta$ by ASO titre (1-2 wks)

Pyoderma $\rightarrow \Delta$ by anti-DNAase B (4-6 wks)

95% PSGN resolves

5% PSGN \rightarrow CKD

Kidney Bx → Endothelial & mesangial cell proliferation & obliteration of capillary lumen.

- Neutrophil infiltration

Immunofluorescence - Granular deposits of IgG & C3
"STARRY SKY"

Good pasture Syndrome has linear deposit of IgG & C3.

Subepithelial humps are a sign of PSGN.

Q Persistently + C3 found in all except:

- a) Post streptococcal GN (normalise after 6-8 weeks)
- b) Mesangio capillary GN
- c) Cryoglobulinemia
- d) SLE
- e) IE
- f) Shunt nephritis
- g) Factor H-mutation → HUS.